The Romberg's Connection 2009 Survey Results

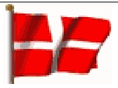
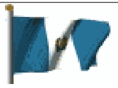
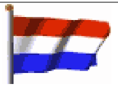
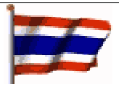
Disclaimer: Please be advised that everyone's experiences may be different and appropriate treatments may vary. Any medical information that you find on The Romberg's Connection website must NOT serve as a substitute for a consultation with one's personal physician. Our visitors should discuss any specific questions or concerns they may have about Rombergs with health care professionals who are familiar with the specifics of their special case. As a support group, The Romberg's Connection is unable to offer medical advice to anyone.

Throughout this survey, when we use the term "Rombergs" or "Romberg" we are referring to: "Parry Romberg", "Parry Rombergs", "Parry Romberg Syndrome" and "PRS".

This is an unscientific survey designed to gather information from those who must deal with Rombergs on a daily basis.

Go to: The Romberg's Connection

These are the countries represented by the (143) people who took part in our 2009 Survey.

 Australia (13)	 Brazil (1)	 Canada (4)	 Denmark (1)
 England (3)	 Germany (5)	 Guatemala (1)	 Ireland (1)
 Israel (2)	 Malaysia (1)	 Netherlands (3)	 New Zealand (2)
 Romania (1)	 South Africa (3)	 Spain (2)	 Sweden (1)
 Thailand (1)	 United Kingdom (8)	 USA (90)	

Number responding = 143 / percent responding = 100% of total respondents (143).

Table of Contents

Introduction

The Romberg's Connection 2009 Survey was released to our members over the Internet on February 17, 2009. The Survey was closed March 11, 2009.

The Survey consisted of two hundred seventeen (217) questions over seven (7) topics. One hundred forty three (143) Connection members provided 14,420 answers to the Survey questions.

A copy of the Romberg's 2009 survey questions can be found here:
The Romberg's Connection 2009 Survey Questions

The purpose of this document is to report the answers provided by our members.

Our thanks go the following people who have contributed questions and/or suggestions to this survey: Dr. Foeldvari, Dr. Stone, Kristina, Markus, Margaret H., Margaret L., Pam, Terri, Lindy and Sally.

Thanks also go to the following for their help in preparing this document: Markus, Katharina, Ray, Brian, Neil and Lindy.

But most of all, our thanks go to the (143) members who took the time to particpate.

For more information about **The Romberg's Connection**
please email us at:
rombergs@hotmail.com

Page 8. I. General Information cont.: Who Is Affected

1. Please select who is affected (Required):

Affected Person	Responses (143)	% of Number responding
Yourself	102	71%
Daughter	21	15%
Son	18	13%
Mother	0	0%
Father	0	0%
Sister	1	0.7%
Brother	0	0%
Aunt	0	0%
Uncle	0	0%
Cousin (Female)	0	0%
Cousin (Male)	0	0%
Granddaughter	0	0%
Grandson	0	0%
Speaking for a Friend	0	0%
Spouse	1	0.7%
Other (Specify)	0	0%

Number responding = 143 / percent responding = 100% of total respondents (143).

Page 6. I. General Information cont.: Person's Gender

1. Gender (Required):

Gender	Responses (143)	% of Number responding
Female	113	79%
Male	30	21%

Number responding = 143 / percent responding = 100% of total respondents (143).

Page 9. I. General Information cont.:
Affected Person Medically Diagnosed and Age?
1. Was the affected person medically diagnosed with Rombergs?

Diagnosed	Responses (142)	% of Number responding
Yes	132	93%
No	10	7%

Number responding = 142 / percent responding = 99% of total respondents (143).

Note: If the answer was "Yes" to the above question 9. I. 1. "Was the affected person medically diagnosed with Rombergs" then the following section (Page 11) would apply.

Page 11. I. General Information cont.: Diagnostic Information
1. At what age was the affected person diagnosed:

Age	Responses (124)	% of Number responding
69	1	1%
57	1	1%
50 - 51	2	2%
45 - 49	6	5%
40 - 44	7	6%
35 - 39	4	3%
30 - 34	8	6%
25 - 29	11	9%
20 - 24	11	9%
15 - 19	20	16%
10 - 14	23	19%
5 - 9	22	18%
1 - 4	8	6%

Number responding = 124 / percent responding = 87% of the total respondents (143).

Page 11. I. General Information cont.: Diagnostic Information

Calculated: Number of Years Until Diagnosed:

Years	Responses (124)	% of Number responding
55 - 59	1	1%
50 - 54	1	1%
45 - 49	5	4%
40 - 44	5	4%
35 - 39	8	6%
30 - 34	4	3%
25 - 29	8	6%
20 - 24	9	7%
15 - 19	4	3%
10 - 14	17	14%
5 - 9	25	20%
1 - 4	23	19%
0	14	11%

Number responding = 124 / percent responding = 87% of total respondents (143).

Page 11. I. General Information cont.: Diagnostic Information

2. Please select the type of specialist who made the diagnosis:

Specialist	Responses (154)	% of Number responding
Acupuncturist	0	0%
Dentist	7	5%
Dermatologist	18	14%
Ear, Nose and Throat	9	7%
General Practitioner	6	5%
Geneticist	3	2%
Neurologist	21	16%
Opthamologist	5	4%
Plastic Surgeon	40	31%
Reconstructive Surgeon	18	14%
Rheumatologist	7	5%
Other (Specify)	20	16%

Number responding = 128 / percent responding = 90% of the total respondents (143).
Note: This was a multiple choice question.

Page 11. I. General Information cont.: Diagnostic Information
2. Please select the type of specialist who made the diagnosis:
Other (Specify)

% of Total Responses	Responses (20)	Other Specialist
1%	2	Pediatrician
0.7%	1	oral surgeon
0.7%	1	immunologist
0.7%	1	Cranial Facial Specialist
0.7%	1	Both Neurologist & Plastic Surgeon
0.7%	1	pathologist
0.7%	1	can't remember! referred from GP to surgeon possibly
0.7%	1	oral and maxillofacial surgery specialist
0.7%	1	Maxo Facial Dr.
0.7%	1	Also visited Mayo Clinic
0.7%	1	One doctor, many hats. He was an otolaryngologist plastic/reconstruction surgeon.
0.7%	1	jaw specialist
0.7%	1	craniofacial surgeon
0.7%	1	eye doctor
0.7%	1	Op Smile Doctor/Plastic Surgeon
0.7%	1	Multiple physicians at a conference at University of California, San Francisco
0.7%	1	oral maxfillio facial surgeon
0.7%	1	physician
0.7%	1	oral surgeon

Page 11. I. General Information cont.: Diagnostic Information

3. Please select the procedure used to make the diagnosis:

Procedure	Responses (192)	% of Number responding
ANA (Antinuclear Antibody test for level of inflammation)	8	6%
Biopsy for bacteria	3	2%
Biopsy for sclerotic content	10	8%
CAT (computed axial tomography (CAT or CT scan))	20	16%
MRI (magnetic resonance imaging)	23	18%
Ultrasound	1	0.8%
Visual Diagnosis	110	87%
Other (Specify)	17	13%

Number responding = 127 / percent responding = 89% of the total respondents (143).
Note: This was a multiple choice question.

Page 11. I. General Information cont.: Diagnostic Information
3. Please select the procedure used to make the diagnosis:
Other (Specify)

% of Total Responses	Responses (17)	Other Procedure
0.5%	1	Dentist took pictures and sent them off for a facial doctor to look at.
0.5%	1	Photos taken
1%	2	Teeth x-rays
0.5%	1	Consult with a team of doctors.
0.5%	1	Experience on past cases
0.5%	1	see below
0.5%	1	Antiphospholipid Syndrome blood panel, Mayo Clinic, infectious disease bloodwork, viruses and bacterial, plus IGM/IGG, Homocysteine Platelet Levels (Genetic), blood clotting, stroke causing test
0.5%	1	Angiogram
0.5%	1	Lots of xrays, bone density
0.5%	1	I don't know
0.5%	1	Biopsy see question 4
0.5%	1	Visual diagnosis after MRI
0.5%	1	Bone scan
0.5%	1	Doctor has another patient
1%	2	X-ray

Page 11. I. General Information cont.: Diagnostic Information
4. Comments for the Affected person's diagnostic information:

Comments (63)
I did not receive informations about the disease, I looked on the internet.
A team of New York dermatologists provided the diagnosis.
After everything else was ruled out (M.S., Lups, Scleroderma, Lyme diseaseothers), noticed an indentation on my forehead. That lead to the diagnosis.
There were numerous neurologic, rheumatic and ophthamology tests.
My initial symptom was en coup de sabre, visually diagnosed by a dermatologist when I was 2 or 3. The phrase Parry Romberg Syndrome was not used until much later, by the plastic surgeon who performed several surgeries and procedures when I was in high school. However, he did not *at that time* call it PRS to me or (as far as they can recall) to my parents. Years later, as a married adult expecting my first child, I had called him to ask him some questions and that was when I first heard the term PRS.
Team of dermatologists (approximately 25) confirmed diagnosis.
"Trigger, looking back appears to have been a knock on the forehead above my left eye during a car accident, at age 14. After that, I began to notice the tissue around my left eye began to disappear around the eye, including the lid area showing more eyelid on the left eye. I also noticed I would rather have photos taken on the left, due to being thinner on that side, under my chin and in the neck area. I had extremely severe headaches beginning at age 15 beginning early Saturday AM after a Friday night football game. I noticed it was the first time my mother gave me 3 adult aspirin at a time. I had severe headaches until June 2002, by then I found out they were migraines. During my teens I had symptoms of stroke, and tias, but didn't know what they were. I had my vision in my right eye cut out on me or become very darkly shaded. Also, a period of bright lights that made me lose vision in both eyes. Upon physical activity, I had a spatial orientation problem on my right side, that did get better playing raqcuetball, but stumbled a great deal over my right toe, while playing, even walking. My left leg is shorter than the right one. Upon review of my Social Security Disability Doctor's review of me, and obtaining a copy of her report, she brought up the stationary Romberg Test. Since many Rombergers are affected by Central Vestibular Dysfunction, basically, interfering in their sense of balance, and spatial orientation. This causes falling at best and neurally mediated or neurocardiogenic syncope (fainting) at worst, or other types of Vertigo, positional or Peripheral. This is basically due to having become more single sensory oriented, while walking forward, with each change in vision, and sense of the height of the step or slope, the brain has to recalculate the height, position, with each step. The other senses necessary to for balance become disconnected causing the Central Vertigo. Due to a TIA with Dysphasia (inability to speak clearly), but able to think clearly in July 2002, I went for an MRI of the brain, that clearly showed the results of years of migraines, as well as old plaques in both hemispheres in the brain. The SSDI Drs.' report wasn't received until May of 2003, and upon review by my ENT, we didn't discover that I had no left cheek muscle. The report from NORD, verified other neurological, myclonic seizures, focal or sensory seizures from plaques in the right brain. The asymmetry was measured, and noted from the face down the body, and under the left rootline of the gum. Audio testing and ENG testing verified Left Peripheral Vertigo, but the Cawthorne's head exercises that should have only taken weeks to cure the problem and put me back on balance, only exacerbated the Vertigo. That is when my ENT realized my gait, floaters in my right eye, and a black spot in my right eye, found while doing the coin toss exercise, and the inability to determine where sound is coming from resulted in the diagnosis of Central Vertigo. Since then, in 2004, I've had another TIA, and an atypical stroke of sorts, with plaques in the gray matter of the brain that the neurologists have missed. Also, in addition to severe clotting factors, my Primary Care and Rheumotologist found a genetic fluctuating platelet elevated level. The Venostat that was conducted in the late 1990s, showed inflammation of the blood vessels, that causes Peripheral Neuropathy (Inflammation and Constriction being an issue for people with Rombergs), but I didn't have the level of Primary Care in Internal Medicine that I have now. So, by looking forward and backward, we've put the pieces together, for the full diagnosis of Progressive Hemi-Facial Atrophy, beginning at around the age of 14, and continuing, with further atrophy until the present, at age 54."
Started as a small quarter size blemish in the affected area - left side check.
I self diagnosed via Internet research more than one year before professional diagnosis.
I have had a couple of 3D Cat Scans which have monitored the progression since I was diagnosed. The blood work came back normal.
It took 8 years to finally find someone who knew what PRS was.
Continued on the next page.

Page 11. I. 4. Comments (continued)
My ENT doctor said that he had heard of Parry-Romberg syndrome, but had never seen a case of it, until he saw me.
In 2001 at the Childrens Hospital in down town Houston, Texas, I was diagnosed by a "Team of Doctors" they specialized in diagnosing and treating children with cleft palates and other child hood deformities caused by some syndrome or illness. I had a CAT scan and X -ray done on my skull. I was asked a few questions by 10 different doctors who practiced different specialties. Finally They told me I had Parry Rombergs and that I was was the 4th person who came in that had Parry Romberge Syndrome.
I don't know exactly how to answer this but, when I was first diagnosed, they called it hemifacial atrophy, then about a year and a half ago, I went to a cosmatologist and he told me the correct name was: Parry Romberg Syndrome, and that it was affecting the right side of my face now. Gave me some kind of cream and told me that there was nothing that he could do for me. So, I haven't been seen by no one else.
The ENT doctor said he had never seen a case of PR syndrome, but had heard about one case.
Very little was given except the effect of wasting of the soft tissue.
I was born with Parry Rombergs, I first went to see a surgeon at the age of 17. I went back to a different surgeon at the age of 37, who gave me a diagnosis on the 3rd visit.
"Diagnosed by biopsy with 'localized scleroderma'. Pediatric dermatologist and pediatric rheumatologist both diagnosed it as PRS. Pediatrician called it Linear Morphea."
Orginally started with hair loss at 15, and later had a reconstructive flap done, which caused some infection and poor healing. Later diagnosed with Linear Scleroderma. Wasn't until after pregnacy that the drastic symptoms came about, diagnosed by Dr. Seibert, for a reconsructive microvascular surgery attempted, which repeatedly got infected probably the first mersa.
Went to GP (General Practitioner) who didnt know what was wrong so referred to plastic surgeon who had seen one case before and diagnosed with Rombergs.
The plastic surgeon took x-rays & photos along with him to a medical convention and came back with my diagnosis as Rombergs.
She also received an MRI and CT scan to establish no neurological effects.
Diagnosed 15 years after surgery to remove affected area on revision of file.
Initially a plastic surgeon said that I had a birth defect when I was examined in my early teens. Then at 16, I was examined be a surgeon at Stanford Medical Center who put my history of seizures together with my facial deformities and diagnosed Rombergs.
Biopsy came back as "localized scleroderma" but all specialists say PRS.
"Have several of the - atrophy of the sucutaneous muscle on half of my face, lip and tongue. - skin overlying affected areas has become darkly pigmented (hyperpigmentation) with, areas of hyperpigmentation and patches of unpigmented skin (vitiligo). - onset of the disease began at the age of 9."
Upon the recommendation of my dentist, I went to eye doctor for difference in pupils (and dents in my head). (I had visual problems since the age of 7). It was very fortunate for me that the eye doctor knew exactly what PRS was.
After getting the diagnosis, we went to a rheumatologist and she did the ANA test and other auto-immune tests.
The specialist (visual) recognized PRS at our first visit to the hospital. We visited the hospital because our son was suffering from headaches.
Half of her face is affected. She was treated by Dr. Thomas Reese in New York University. She still suffers with the condition. It was first diagnosed as Progressive Hemi Atrophy.
There were physical tests involving electrodes, there was some system for ruling out scleroderma, but it was so long ago that I don't remember. Don't have the records.
Plastic Surgery
Took some time for diagnosis and was made by a team of doctors at Children's Hospital.
My PRS showed already as a child - but I did not ask for diagnosis until I was 32.
Continued on the next page.

Page 11. I. 4. Comments (continued)
Was first diagnosed with Lupus (later found out that was an incorrect diagnosis). Was then refered to reconstructive surgeon who made positve diagnosis of PRS.
"Facial asymmetry was first noticed by orthodonist (though did not know PRS), then primary care physician also noticed (though did not know PRS) and she referred us to craniofacial specialist at Children's Hospital in Boston where son was diagnosed. This was confirmed at Massachusetts General Hospital, also in Boston. I was unaware of other types of procedures to diagnose PRS."
It took a long time as only the eye doctor knew what it was. Then I was referred to a plastic surgeon.
"On the left side of my face I had: coup de sabre droopy eye lid cleft in chin hollow cheek droopy lip."
"I had a CT Scan, 2 MRIs of the brain, EMG, and multiple blood tests to rule out other diagnoses. Finally saw both a neurologist and plastic surgeon at the University of Pennsylvania in the fall of 2007 and both diagnosed Parry-Romberg."
I went to see my GP concerned that one side of my face was slightly swollen. My GP thought it was the other way around, that one side of my face had started to atrophy. He then went away to research what it might be, which was when he found out about Rombergs. I then had an MRI to see whether the muscles and/or soft tissues of my head had been affected.
Dermatologist + Neurologist in cooperation (University Clinic)
It took two years for the condition to present itself in a form where the diagnosis could be made. Prior to that, it ranged from "nothing serious" to mast cell disease, dermatomyositis, appearance of Sturge-Webber, AVM.
I was referred by my regular doctor to a group of plastic surgery doctors.
"Atrophy to the L hand side of the face, trunk, and back. Hyperpigmentation on affected areas."
Dr. Mintz ran a lot of test, including bone density and a special x-ray that took an hour with radioactive dye that I don't kow the name of prior to surgery.
The only symptom I had was a 7th cranial nerve palsy (Bell's palsy) the investigations to find the cause of this were CT and MRI scans. PRS was diagnosed off brain findings on MRI.
"Ten years between onset and diagnosis age 24 onset age 34 reconstruction surgery."
1. first saw dentist. She realized something was not right. She thought she was developing a cleft lip but knew that cannot happen. She referred us to see a plastic surgeon and also asked if it was ok to send her teeth x-rays to other doctors. Plastic surgeon diagnosed her and then ordered a CT scan. He then referred us to rheumatology and he further diagnosed her with the scleroderma. And he referred us to a genetic doctor, mris, eegs, neurology, audiology, and an allergist. And also a counselor.
Depression and lost of fat / muscle at affected area.
Blood tests did not confirm the diagnosis however, we were told that this is probably an onset of PRS.
Left side of the face, cheek, chin, lip. Not the forehead.
Dr. Grillion just said it.
Visited Dr. Kenneth E. Salyer in Dallas after learning about PRS and he confirmed the self-diagnosis. Didn't recommend surgery as the case at the time, was relatively "minor."
My diagnosis was made by an ear-nose-throat plastic/reconstructive surgeon. He took one look at me and told us what I have. He's worked on PRS patients before.
Diagnosed with PR at age 29 by dentist (TMJ specialist). Afflicted with PR since around age 11. Diagnosed with scleroderma (not linear) at age 35 by rheumatologist (subsequent to first pregnancy). Likely had scleroderma for many years as well, however, both PR and scleroderma flared immediately following pregnancy.

Continued on the next page.

Page 11. I. 4. Comments (continued)
I was only 15 years old when they did my biospy. I do not know what type it was but they removed two sections of my skull the size of a quarter.
Immediately after giving birth to my son at age 23, my left side of my face went flat. I saw a dermatologist who referred me to a plastic surgeon.
Visual diagnosis was also used - Neurologist looked at pictures of me at an earlier age. He was also willing to look at the information I brought from the NORD site. It's important to get a doctor who will listen and also do a bit of research. Dermatologist diagnosed me with another disease, that I as a lay person knew enough about to know that it was inaccurate - she was very dismissive.
"ANA originally negative; a year later, ranged between 80/speckled, 40/speckled, 160/speckled Doctors go back and forth--say it looks like LS but acts like PRS--elements of both Say mild so far, but have seen changes over 3 years, so don't know what "mild" means; is it just starting? How do you know if it will stop? Child also showing signs at same time"
66
Don't really know much about diagnosis as I was a child- can remember having EEG, ECG and being "on display" for medical students in the public hospital system- not good memories.
Initial diagnosis by plastic surgeon was that of "hemifacial atrophy." Follow up exams at Mayo, but do not recall a specific reference to "Rombergs." One endocrinologist there mentioned linear scleroderma; when I asked about that, she said it was the same thing.
I was referred to Dr. Joseph Murray at Peter Bent Brigham Hospital (now Brigham and Women's) in1973 by a Plastic Surgeon. Dr. Murray diagnosed me, and told me that there were only 50 known cases in the country.

Number responding = 63 / percent responding = 45% of total respondents (143).

Page 9. I. General Information cont.:
Affected Person Medically Diagnosed and Age?
2. Was the affected person medically diagnosed with Linear Scleroderma?

Diagnosed	Responses (138)	% of Number responding
Yes	37	27%
No	101	73%

Number responding = 138 / percent responding = 96% of total respondents (143).

Page 9. I. General Information cont.:
Affected Person Medically Diagnosed and Age?
3. Which diagnosis came first?

Came First	Responses (117)	% of Number responding
Linear Scleroderma	24	21%
Rombergs	93	79%

Number responding = 117 / percent responding = 82% of total respondents (143).

Page 9. I. General Information cont.:
Affected Person Medically Diagnosed and Age?
4. What is the age of the affected person in number of years?

Age	Responses (143)	% of Number responding
70 - 74	3	2%
65 - 69	3	2%
60 - 64	4	3%
55 - 59	10	7%
50 - 54	14	10%
45 - 49	19	13%
40 - 44	8	6%
35 - 39	17	12%
30 - 34	10	7%
25 - 29	13	9%
20 - 24	4	3%
15 - 19	13	9%
10 - 14	11	8%
6 - 9	13	9%
1 - 5	1	1%

Number responding = 143 / percent responding = 100% of total respondents (143).

Page 12. II. Rombergs Symptoms History
1. History of symptoms:
Age of the affected person when the symptoms started?

Age	Responses (137)	% of Number responding
67	1	0.7%
60 - 64	1	0.7%
50 - 55	1	0.7%
45 - 49	1	0.7%
40 - 44	2	1.5%
35 - 39	1	0.7%
30 - 34	3	3%
25 - 29	4	3%
20 - 24	7	5%
15 - 19	10	7%
10 - 14	31	23%
5 - 9	37	27%
1 - 4	29	21%
At Birth	4	3%
Unknown	5	4%

Number responding = 137 / percent responding = 96% of total respondents (143).

Page 12. II. Rombergs Symptoms History

2. Are the symptoms still active?

Active	Responses (132)	% of Number responding
Yes	82	62%
No	50	38%

Number responding = 132 / percent responding = 92% of total respondents (143).

Note: If the answer was "Yes" to the above question 12. II. 2. "Are the symptoms still active?" then the following section (Page 13) would apply.

Page 13. II. Rombergs Symptoms History cont.: Symptoms Still Active

1. Comments for the symptoms still being active:

Comments (56)
I think that the hole in my face it gets bigger and the part of the face that is affected it's getting bigger.
My symptoms appear to be slowly active in respect to the slow deterioration of the health of my eye, in terms of the vision, and the dryness of the cornea and double vision. All are caused by Parry-Romberg.
In looking at my optic nerve, we can see that the disease is still progressing. I can't tell cosmetically if it is or isn't.
I have seen through photos the affected area getting worse. Expecially in my late 30's it started off very severe.
"Hard to see the changes when we are with our son every day, does not seem to be progressing rapidly."
My left side continues to atrophy. It is obvious especially when a new dip develops (next to left ear). The 3D Cat Scan also shows progression. I have a couple of scans to compare the progession to. My dental x-rays also show another tooth with atrophy as well as the bone disappearing. The gum recession is obvious visually.
Muscle spasm on face and jaw area
Surgery needs repeating due to fat graft in the left cheek reducing and the eye continues to become more recessed.
We are not sure if the PRS stopped or reversed, although it seems it is reversing. No longer have symptoms of facial pain and numbness since treatment.
"I still have a lot of itching on my right side of face. Strange sensations on top right side of my head and right forehead. I have noticed that the right side of my face has atrophied somewhat more."
THE "BALD SPOT CAUSED BY THE EN COUP DE SABRE IS MOVING TOWARD THE BACK OF MY HEAD. IN THE BEGINNING IT WAS ABOUT ONE INCH BEYOND MY FOREHEAD. NOW IT IS ABOUT THREE INCHES.
My face itches a lot when it is active and feels like it is next to heat.
Stress and/or low immunities (general run down of health) affects whether the Rombergs becomes more active.
As far as I can tell, my symptoms have never stopped since I was first diagnosed. My most telling symptom, besides the atrophy, is itching by my right temple.
Ringing in the ears...twitching around graft, dizziness/vertigo.
Brown streaks running from his cheek to his neck. A plaque-like scar on his cheek, where the atrophy is. Doctors said his tongue has a little atrophy too.
We are not completely sure but it seems to be deeper and one side of her forehead is lower than the other, seems to change and she still gets headaches.

Continued on the next page.

Page 13. II. 1. Comments (continued)
My face does not seem to be getting any worse but I am still losing soft tissue on my foot.
I believe I have noticed a progression in the symptons of atrophy in my forehead over the last few years. I also have continued to have jaw difficulties that I believe are related to PRS. Finally, the headaches and occasional migraine that I have experienced continue.
You learn to live with them.
Electrical impulses in my left temple. Left eye socket seems to be sinking further into my head. Legally blind in left eye, last exam vision in left eye was 20/1100.
Over the years the Rombergs area on my cheek has developed a long hard indent. My eyesight in my left eye has deteriorated much faster then my right eye.
Seems to be gradually still getting worse.
Atrophy possibly got worse during first pregnancy. Other symptoms such as SPD / arthritis type symptoms during second pregnancy, but obviously impossible to say with certainty whether these are PRS related. Left and right pupils different sizes, likely to be PRS related but again cannot be certain.
Ooop- I wrote that on the last part- sorry!
Continue to have progressive vision loss and tissue loss. Very slow. Stops and starts again.
Yes
Not sure, we think its slowed down or stopped.
As mentioned before, headaches still present, but tissue disappearance stopped/stable.
"Indentation in the forehead in the shape of a V Several bald patches on top of the head Recessed eye, vision affected Indentation on the chin"
Had surgery in 1992 & 1995. PRS has been dormant for many years but I'm starting to see the atrophy start up again.
"I still become smaller all over my right side of the head - not 'only' my face, but also on the top of my head. I have trigeminal nerve pain in affected side. It feels like far inside of my ear. I do not know if my brain or blood pressure is affected. My blood pressure is quite low - in the perspective that I have 20 kilo over weight and smoke 30 cigarettes every day."
Still loss of eye lashes, eye ache from time to time, some re absorption.
"The cleft on my chin, my droopy eye lid and skin discolouration on my forehead and neck have worsened over the last 2 years. I've also developed a small dent above my left eye brow over the last year. When I lose weight, it happens more dramatically on my left side, particularly from my face and breast. I even have less of a 'roll' around my middle on the left side. When I had a bad flu about 18 months ago, my temperature was higher on my non-PRS side (I think about 2 degrees Celcius)."
Migraine headaches, seizures (Grand Mal) joint pain, continued wasting of parts of the face, especially under the affected eye. Her own tissue injected into that area has been reabsorbed although most of the free flap has remained healthy (cheek area).
My natural weak tissue still is disappearing. My right side gets skinnier in time. Also does my skin change color, from normal to red in an expanding area and it gets more intense red. And my eye started shedding tears and became very sensitive.
Ten years on very very slow progression, with periods of no progression. Illness, stress, tiredness seem to bring it on and my face gets very tingly especially in the areas at the end of the worst areas. At present it is my lips and inside my mouth that seem to be the more active areas.
Hard to tell if it is, or when it stopped and started again.
Plastic surgeon commented last fall that the left side of my face was affected. That had not been seen before.
It has been a gradual progression for around 10 years, but it is hard to say whether it is still active or not as it is so gradual. I have had 3 fat injection treatments, which also makes it hard to tell.
Bruising like markings have gotten slightly bigger. And more noticeable on nose and around left eye.
Continued on the next page.

Page 13. II. 1. Comments (continued)
"I am currently 29 this is happening to me now!!!! These symptoms may be progressing at this point in time: mild weakness in left arm and gradual loss of pain sensation in left leg."
The atrophy on the right side of my face is very slowly getting worse. I do get pain through my right eye and some neuralgia on the affected side, but it's not too bad really.
Degradation of facial symmetry continues in a very slow fashion.
My symptoms started only in the last year, and are progressing quickly on my face.
"The PRS seems to have accelerated in the last decade. It is now migrating down my neck and I have a sunken spot in my skull (but still hair there). I lost hair over my affected ear and almost all muscle tone there. I'm starting to lose muscle around my eye and more around my mouth. I chew my lips sometimes when chewing. And my jaw is starting to pop and grind, but without pain. I'm currently taking Prednisone and Methotrexate and the pain in the jaw area has been eliminated, though the deterioration still continues."
I now am still experiencing twitches in my right cheek, macular degeneration on my right eye, just had that operated on 4 months ago, I am also getting headaches again and have poly neuropathy which my neurologists claim has come from the Rhomberg's.
Symptoms agressively active currently.
"Atrophy still occurring, continual eye problems, have double vision looking in a certain direction, have flashing lights, have dry eyes with blurred vision, have balance problems."
Atrophy on right side of his throat /neck.
11
Have bruised sensation from time to time in chin area, where changes are.
Symptoms are not still active.
The same symptoms as reported.
There appears to have been some further progression in the last five to eight years - the left eye seems deeper (more difficult to insert a contact lens on that side), and there also are further changes along the left side of my nose and chin. Perhaps unrelated, about two years ago, following a severe headache, I noticed my pupils were different sizes, which continues to this day.
Tissue wasting away, also diagnosed with hypothyroidism - hair loss not sure which one is causing that. Have headaches.

Number responding = 56 / percent responding = 39% of total respondents (143).

Note: If the answer was "No" to the above question 12. II. 2. "Are the symptoms still active?" then the following section (Page 15) would apply.

Page 15. II. Rombergs Symptoms History cont.: Symptoms Are Not Active

1. At what age did the symptoms appear to become inactive?

Age	Responses (49)	% of Number responding
45 - 49	1	🟥 2%
40 - 44	1	🟥 2%
35 - 39	3	🟦 6%
30 - 34	2	🟥 4%
25 - 29	3	🟦 6%
20 - 24	9	🟥 18%
15 - 19	9	🟦 18%
10 - 14	9	🟥 18%
5 - 9	6	🟦 12%
1 - 4	1	🟥 2%
Not Sure	5	🟥 10%

Number responding = 49 / percent responding = 34% of total respondents (143).

Page 12. II. Rombergs Symptoms History

3. Have the symptoms ever stopped and then started again?

Stopped Started	Responses (133)	% of Number responding
Yes	55	🟥 41%
No	78	🟦 59%

Number responding = 133 / percent responding = 93% of total respondents (143).

Note: If the answer was "Yes" to the above question 12. II. 3. "Have the symptoms ever stopped and then started again?" then the following section (Page 14) would apply.

Page 14. II. Rombergs Symptoms History cont.: Symptoms Reactivated

1. Please comment on why you think that the symptoms were reactivated:

Comments (45)
I don't know .....I had a period around16 when it stopped for like 6-8 months, the skin was not colored, the pain stopped but after that it started to ache and then followed it's course.
See explanation of #13.
I got into a car accident when I was 17 years old. I sustained a laceration above my right (affected) eye. The forehead atrophy and subsequent eye involvement seems to have started after that.
As noted previously, I'm not completely certain that I've had a reactivation, but if it was a PRS reactivation that caused the fat grafts in my chin to begin atrophying, then it would likely have begun during pregnancies with my oldest two children.
At the age of 4, a dark mark occured on my cheek, during my 30's the dent started in my cheek and forhead.
The fat injections were absorbed on left cheek and lip.
stress
I think my symptoms got reactivated due to pregnancy issues and hormones.
I'M GUESSING THAT PROBLEMS WITH ANOTHER AUTOIMMUNE DISEASE (POLYMYALGIA RHEUMATICA) MIGHT HAVE CONTRIBUTED.
I had no sign of PRS on the right side of my face until I had went through a stressful time with my daughter, because she has systemic lupus arethamatosis with nephritis, then about the year of 2007, I noticed the right side of my face starting to have like a dent about the size of a dime. I had hoped that it wasn't that, but it was.
Teenage stress over appearance and comments from others.
Stress in adulthood, general development throughout the lifespan that we all encounter...this just has its own set of issues...
No idea maybe stress plays a part in it.
Can see changes in the face and eye.
"Childbirth for certain. Then was doing quite well until a car accident in which the blunt trauma led to seizure activity. Seizures continued for 3 years until ultimately they took her life."
Again it is my foot that seemed to stabilize and then started to get worse again recently.
I wish I knew why. The symptoms seemed to be dormant for the last 20 years and in the last 18 months I've notived my left eye seems to be sinking more into the head.

Continued on the next page.

Page 14. II. 1. Comments (continued)
Pregnancy
No idea. I did get pregnant twice over 10 years ago but cannot say Rombergs was re-triggered by that. I do not put it down to aging as my normal cheek has not indented & there is not much wrong with my right eye sight.
I definitely think that lately it's been because of stress! I also feel that having been pregnant in the past (I have 2 children) has caused this disease to be active, but I also have Systemic Lupus Erythematosus.... so, having two rare and odd diseases, its hard to tell!
"- irritating tingling - the tissue seemed tighter"
After each pregnancy and again, more recently, with menopause.
?
Because I lost a lot of weight and when that happens I seen a dramatic change in my face.
Not sure
menopause?
Possibly related to pregnancy or stress.
I'm not sure, maybe hormonal changes. (I am now what is know as middle age) ;-)
Don't know
"The coup de sabre came on when I was pregnant with my second child when I was 26. It didn't go away after the birth. About 6 months after the birth, I had eye surgery (for a squint) and started to notice the difference between the left and right sides of my face. I didn't ask anyone about it at that time because everyone says that no one's face is completely symmetrical! When I was 29, I lost almost 10 kilos in over about 6 months. That's when the dramatic difference in my face became noticeable enough for me to ask my doctor about it. My left cheek was hollow and the cleft in my chin became more obvious. I also asked about the coup de sabre at that time (it hadn't gone away since the pregnancy)."
Age 23, neuroneuritis and vision loss in left eye. Syptoms stopped at age 12 and then this occurred.
I have no idea. Everything seemed to have stopped for a few years, and due to so many operations you could hardly see I have Rombergs anymore, until a few years ago.
Had surgery in May of 89 and November of 89...lost most of the grafts that were put in due to body rejections but noticed that in 90 my face had started to atropy again.
pregnancy
I don't know if they were reactivated. I cannot comment.
Unsure on this.
Going off of the birth control pill, therefore being subject to the natural monthly hormone fluctuations, then having four children and all the hormone swings that brings.
I honestly have no idea as to what caused this to act up again. It has been more than 26 years since it has stirred up again.
"1. Life situations involving extreme, long-term stress. 2. Pregnancy"
I'm wondering if it was the stress brought on when my child began to be tested for pigment changes. At first they thought he had an endocrine issue, but now it appears we are in the same boat. Stress has been daily and relentless. Am working on managing it daily.
.... ????????????? ....
Symptoms reactivated with each pregnancy then deactivated.
Continued on the next page.

Page 14. II. 1. Comments (continued)
Stess, anxiety, depression.
Pregnancy reactivated or at least exacerbated the symptoms. Not sure what could have spurred things on in the last 5 to 8 years. Possibly stress.
The symptoms that I have had seem to be a progression, not a reactivation.

Number responding = 45 / percent responding = 31% of total respondents (143).

Page 12. II. Rombergs Symptoms History

4. Is the person facially affected on:

Facially Affected	Responses (127)	% of Number responding
Left Side	68	50%
Right Side	64	47%
Both Sides	5	4%
Not Affected	0	0%

Number responding = 137 / percent responding = 96% of total respondents (143).

Page 12. II. Rombergs Symptoms History

5. Where on the affected person's body do the symptoms also appear:

Person's Body	Responses (129)	% of Number responding
Left Side	30	23%
Right Side	23	18%
Both Sides	8	6%
Not Affected	68	53%

Number responding = 129 / percent responding = 90% of total respondents (143).

Page 12. II. Rombergs Symptoms History
6. Comments for Rombergs Symptoms History:

Comments (65)
Symptoms started as "shortening" of my right jaw/chin at age 24- around age 37-38 tissue started to go away and one indentation started on my cheek bone.
I did not know anything was wrong until I lost vision in one eye at age 25. They tested for everything and it wasn't until I noticed atrophy on my forehead (above affected eye) that I got a diagnosis. Since then, it appears that my jawline and nose are also affected. I am not sure if I didn't notice, or if it reactivated in my twenties/thirties.
PRS affected my left arm/hand & left leg/foot.
In the past 10 or 15 years I have had marked atrophy of the chin area where fat grafts were done 30+ years ago. I am uncertain as to whether this is active PRS or simply the loss of the fat grafts over time. I have not observed any new areas of atrophy.
The left side of my face is affected. Whole Left skull bones in CAT scan. Left jaw, lips and chin in soft tissue, (muscle and fat) and connective tissue have atrophied. Nose is turned slightly to the left. Eye socket sits lower then the right. Left ear sits lower then the left ear canal is small and narrow. Occiput hole in back of skull is smaller on left then right. Base of skull is turned to the left on axis. Brown discoloration on left neck, and jaw with some blood vessels that are superficial. Brown discoloration on right side of back with indentions also has blood vessels that are superficial.
Some time in the late 1990's, I had two minor traffic accidents about two or three years apart. My car was rear ended on both of those occasions at very slow speeds (I was stopped both times, and the car(s) behind me hit my car). I had some right neck pain and have wondered if that was what might have triggered my Rombergs. I have also noticed that I have a lot of itching (off and on) on the right side of my forehead and my right ear. I feel the itching has something to do with the Romberg's as there is no itching on the left side of my face. I also noticed about 8 or 9 years ago that my right eyelid drooped and eventually, I noticed that my left cheek was fuller than my right cheek and mistakenly thought that I had a sinus infection in my left (good) cheek). My General Practitoner told me that I didn't have a sinus infection, but referred me to an ENT doctor. My ENT doctor ordered a CAT scan and then diagnosed me with Parry-Rombergs syndrome. He stated that he had heard of this syndrome, but had never seen a case of it.
White spot on lower left chin area . Slower eruption of teeth on left side. Smaller teeth, atropthy of tongue.
I had a cousin who also had it, but I didn't get to meet her, I wanted to, but she passed away before I could.
Symptoms began at age 8 with a nickel-sized lesion on the top, lefthand side of the scalp which was itchy and then became bald and indented. Next at age 11 came another itchy lesion above the left eyebrow. The skin darkened slightly and formed a vertical indentation. Then came around age 15 a slightly noticeable wasting on the left side, forming a perfect line from the outer edge of left eye down to just above the chin. This wasting was not truly noticeable until about age 20, as I was fully mature and had thinned out from "baby fat." I had not linked all these events until a "Eureeka!" moment at age 20.
Tingling and pain on the affected side. Pain in the jaw, ears, head, pins and needles in hands, numbness arms and legs, near blindness in the affected eye, eye developed a squint, double vision, went smaller then the other eye. Sharp pains in the eye like a needle sticking in to it. Mouth affected, deep hole in the upper pallet. So many things over the years I have forgotton about, until someone on the group mentions them.
Started with a stain on the belly.
I am in the beginning of menopause and I suspect that I may have symptoms starting again, but it's so subtle, I'm not sure yet.
It started like a rash under my eye then i had it under my chin and at the corner of my right eye near the hair. during it's activities i had headaches problems eating, it was colored ....like a bruise and very hard until the end of the atrophy.

Continued on the next page.

Page 12. II. 6. Comments (continued)
First symptom en coupe de sabre, which goes from crown to chin, loss of eye brow and lash hair, later fat loss.
Looking at photos, I believe PRS started when I was a teen. However, I didn't notice anything until I had my wisdom teeth pulled at 24. I felt the "hole" in my cheek was related to missing my wisdom teeth on my left side. In my mid 30s I questioned my new dentist about it and he told me that the wisdom teeth had nothing to do with it and I should talk to my doctor about it. That started me down the path of discovery that I had PRS.
A small white mark appeared on the left side of my temple next to my eye. This grew and over a few years turned medium brown & the skin was shiny. Areas affected (all on the left) are two marks on upper lip, large mark covering most of my left cheek. A long paler mark just in front of my left ear. A large mark on my back. All the areas lost the tissue/fat under the skin. Facial bone development was stunted and my eye sockets are not level with each other. Teeth are extremely misaligned.
In June 2008 son was diagnosed; in Dec 2008 he underwent microsurgery with Dr Siebert at New York University. So in reference to Question 2 & 3 above, it is impossible at this time to determine if symptoms are still active. A second surgery in scheduled for 4/09.
Two good hits to the head (one in front and one in back) approx age of 7. Shortly after had fainting spells and the start of visual problems.
I was born with a brown sign on my left cheek. I can't really remember but the atrophy started at age 5-7?
First symptoms were just discoloration. After 2 years of that, the atrophy started.
Gray hair age 5 Pigment changes, migraines age 7-8 Subtle facial atrophy age 9-11 Progressive wasting of soft tissue and fat, tongue, lips Age 17 moved jaws surgically to realign
The first manifestation of the disease was that I had grand mal seizures at the age of 9. My facial deformities weren't apparent until a year or so later.
It is hard to say when the symptoms started but looking back at pictures, I believe it was in my mid 30's. That is also the same time that I lost several teeth on my affected side. At the time, we had no idea how my roots could have atrophied so quickly. I had just seen the dentist 6 months before.
No pain symtoms.
Lots of headaches in childhood, with 'sinus' headaches increasing until adulthood. Sensitivity to heat..profuse sweating...sick to stomach. En Coup de Sabre is not noticable, but can be felt through skin, extending to flat spot on crown. Gray streak in hair appeared at 7 y/o. SX's at birth included a bruising on lower back, just above hips--noticed by paternal gm when changing diaper. "Wasting of face" became prominent in early 20's with burn continuing through 30's.
The first thing that my barber noticed was the indent in my skull. He told me that he saw other patrons with similar indents and it was caused by using forceps during delivery. About a month or 2 later, I started getting headaches and went to see a neurologist. Without doing any tests, he told my parents that I had the classic "coupe de sabre" and was sure it was Rhomberg's Syndrome. He was the doctor who performed the biopsies.
Unsure if the symptoms are getting worse or not.
19 - 22yrs gradual onset of 7th cranial nerve palsy on right side of face 25-26 yrs gradual onset of moderate left leg weakness and loss of balance, occasional vertigo. TWO slightly pigmented, indented lines running down forehead appeared 27 yrs mild weakness in left arm 28 yrs ONE mildly pigmented, indented line appeared runing down nose from eyebrow level to tip of nose. 29 yrs gradual loss of pain sensation in left leg
Hair loss, then indention(s), largest one is down her forehead to between her eyes mostly to the left.
Symptoms seemed "dormant" or very slow in progression from age 10 -16. Then ages 16-18 visible progression in short amounts of time.

Continued on the next page.

Page 12. II. 6. Comments (continued)
She has markings which look like bruises on her forehead, left eye, dark rings around both eyes, down her nose, and under left side of nose, on upper and lower lip on left side, and brown markings from her left side of face all the way in one line to her chin along her jaw.
Left side of face, especially forehead and orbit. Significant progression following each pregnancy, primarily causing the eye to not close properly when sleeping, leading to corneal dryness. Right ear and right side of neck occurred much later - probably late thirties. The skin is extremely thin on the neck. The right earlobe is gone and skin very thin over the firmer cartilage.
My right ear is smaller than my left ear.
Left side affected most notably the face, eye. Also left breast is much larger than the right, indentation on my back between shoulder blades that is very sensitive. Also having episodes of what feels like electricity going through my left temple. Doesn't happen constantly, however I would have maybe one episode every few months and now it seems to happen every couple of weeks.
Started on the lip in middle of face first specialist thought it was frostbite way back then. (it doesnt even snow here). Dentist was the one who picked it up as area appeared to be "growing" referred to an oral specialist who decided to do investigative surgery (lost two front teeth) as it affected gum, bone and lip. By the time of the surgery, it had advanced quickly and affected nose. Following surgery there was no advancement. Suffered severe migraines but that was put down as related to something else. One side of face slightly lower than other but all else ok.
Parry-Romberg has effected the skin, tissue, muscle and bone on the left side of my face. Also effected has been been my eye muscles, eye lashes, cornea, and sclera. I had a spontaneous filtering bleb in my sclera which required a graft to cover.
En coup de sabre at age 8 progressing through to age 21. Chin and jaw midteens through age 24 Right cheek age 20 Symptoms appeared on forehead approximately 2 months after trauma to mid forehead.
Had an impact injury 2 cm round at the top of my forehead. Rombergs started from there. Just before the incident my four children all had chicken pox and I had been suffereing from a virus which was similar to glandular fever or Ross River fever. It took months to get over the virus and in those first six months the progression was rapid but has slowed over the past ten years. Doctor thinks my body was fighting the virus and then got confused by the injury which trigger my immune system to attack the injury site.
Symptoms started with white patchs around jaw line which then progressed with loss of soft tissue, a dent at the top of the head on right side and a small indent on forehead, hairline.
Seizures, starting at the age of two and a half and en coup de sabre at the same time. Gradually the en coup de sabre became an atrohy of the left side of the face. Till today he suffers from epileptic attacks. Only in the last year he suffers from over 200 seizures.
Question 2 "Are the symptoms stll active" should have maybe option. I think the symptoms are still active but have slowed down.
Before age 5: White spot on neck, gray hair, missing eyelashes, asymmetrical nose Additional facial changes apparent at about age 9. Disfigurement quickly became apparent from age 9-17. Surgical intervention began age 16 with good results.
The whole left side of my body is noticably smaller than the right. My brain is the same size on both sides, however the electrical signals from the RIGHT side of my brain are lower (tested by "Transcranial Magnetic Stimulation"). It has not been determined if this is linked to my PRS or not, as this result is apparently somewhat unusual.
I was born with left side facial paralysis (just could not independently move the left side of my face) or it may have happened in early childhood - my first memory is about 6 when I could not wink with my left eye - this was diagnosed at age 54/55 as possible mild cerebal palsy due to premature birth, but having later been diagnosed with PR, I wonder if this was the start of it but on the other hand it could be two different conditions.

Continued on the next page.

Page 12. II. 6. Comments (continued)
The right side of my face is definitely affected. My plastic surgeon also sees signs on the left side of my face. The right side of my tongue is also affected.
It first presented on the face as a violaceous rash. In all the places where the redness or rash was present are now the areas of atrophy.
Gradual hemi-facial atrophy was noticed first; small spots of vitiligo on the head (which has made the nearby hair lose pigment); migraines assumed linked to Rombergs; more recently a persistent case of episcleritis (inflammation of one part of the eye), assumed because of the fact that my eyelid on the affected side of my face does not always close properly when sleeping, causing the eye to become dry and susceptible to irritations.
Have forehead line, line under chin and another brown area on other side of chin. Right side has had nerve (trigeminal) sensations on and off--that's what originally made me sit up and take notice (that and a photograph) Left thigh has an indentation in it. Changes evolved as follows: pigment blotches, face pain, forehead line and small indent, left thigh pain and indent, line on chin (right side), line under chin indents a bit, now brown spot on left side under chin. Cheek changes as well, but doc says does not know if that's age or condition. Have had body-wide fasciculations for past couple of years, don't know if that's attributable to medication or what? Had lyme in 2004.
Suffered jaw spasms and migraine - not for last 10 years or so though.
My symptoms first started out as being a darkening of skin near the lower corner of the left side of my mouth- people used to always comment that my face was dirty. My nostril openings were always different shaped too, and I've always had a "dent" in the top right side of my head as well as a flat and bald spot on the back of my head as well. Although this disease has started and stopped over the years, I've noticed lately that it has seemed to have ramped up again in my face on the left side as I can feel something odd there as well as pain around my eye. I've also suffered a Grand Mal seizure for the first time in Oct. '08 which has been attributed to 3 lesions on my brain. I believe these are on the left side- opposite from the en coup de sabre- weird! The en coup de sabre has run down into my right eye brow which is pretty much gone now too. Luckily, none of the rest of my body seems to be affected at this point though!! :)
At age 5, a rash appeared on the left side of his face. (cheek) For the next 2 years, it changed colors from red, to pink, blue, white and then a mocha-brown color. In that time doctors said it was an allergy, another doctor said it was Atopic Dermatitis. A few months after his 7th birthday, we first noticed the atrophy although we didn't know what we were seeing, he just looked kind of "puny". About 6 weeks later the atrophy was obvious and we rushed him to the doctor with 3 years worth of photos and got the diagnosis. He has atrophy on cheek and a little atrophy on his tongue.
Face affected (above right eye, nose little bit) from age 5 until 7, after that stopped and stayed stable. Headaches started at age 4 and still suffering from headaches (some periods often for 4 days a week, some periods without headaches for several weeks).
Hearing loss on the affected side, as well as facial hair growth.
Tightness in the neck and back muscles of the left side.
Affects my face and my foot on the right side. Have lost soft tissue from the base of my foot - sole and heel.
PRS is contained to the right side of my face. Was extremely active after pregnancies. (age 18-21)
We are not sure if it is still active. I had trouble answering question #2 above.
I am not sure that she has problems externally, but has many problems believed related to Rombergs.
At the age of 12, the skin around the right side of my mouth started to droop.
When I was approximately 9 years old, I had facial trauma that left a bruise on my right jawline. The bruise never went away, and the tissue on that spot thinned. At 14, the hemi-facial atrophy happened seemingly overnight. Dr. Murray injected 11 cc. liquid silicone in my cheek and jaw at 21. In my late 20's, I lost tissue around my right eye. Atrophy began in optic nerve of affected eye at 30. Grand Mal seizures in 40's controlled well by medication.

Continued on the next page.

Page 12. II. 6. Comments (continued)
The questions about whether its still active or not..........I'm not sure.
I did not know some of my sytoms until i read your web site and a lot made sense. Loss of eye lashes and moustache, jaw spasms, eye ache, dent in scalp, then over years loss of fat in eye, face, nose and chin.
The only area affected aside from the face is a small section on her back (same side) where soft tissue/muscle seems to be missing.
I feel that the major progress in symptoms happened between 18 and 28. Since then the progression is very very slow but has not stopped altogether.
Started with a dark line and progressed to the left side. Also affected her teeth and gums on that side. Some pain and weakness on left side later down the road. Did not answer #2. Patient is now deceased.

Number responding = 65 / percent responding = 45% of total respondents (143).

Page 16. III. Associated Symptoms of Rombergs (page 1 of 4)
1. Please select all that apply (page 1):

Symptoms	Responses (420)	% of Number responding
Abnormal sensation in affected area	65	48%
Atrophy (tissue loss)	131	96%
Blood pressure	10	7%
Bone loss	66	48%
Brain affected	27	20%
Chin affected	75	55%
Ear affected	46	34%

Number responding = 136 / percent responding = 95% of total respondents (143).
Note: This was a multiple choice question.

Page 17. III. Associated Symptoms (page 1 of 4) cont.: Abnormal Sensation
1. Please select any abnormal sensations in the affected area:

Age	Responses (134)	% of Number responding
Burning	14	22%
Coldness	16	25%
Pain	35	56%
Tingling	47	75%
Other (Specify)	22	35%

Number responding = 63 / percent responding = 44% of total respondents (143).
Note: This was a multiple choice question.

Page 17. III. Associated Symptoms (page 1 of 4) cont.: Abnormal Sensation
1. Please select any abnormal sensations in the affected area:
Other (Specify)

% of Total Responses	Responses (22)	Other - Abnormal Sensation
0.7%	1	dormant
0.7%	1	glowing skin, no hair
4.5%	6	itching
0.7%	1	muscle cramping
0.7%	1	No feeling when weather very cold.
0.7%	1	not sure
3.0%	4	numb
0.7%	1	pressure
0.7%	1	skin thinness
0.7%	1	slight numbness
0.7%	1	tightness
1.5%	2	twitching
0.7%	1	when it's cold

Page 17. III. Associated Symptoms (page 1 of 4) cont.: Abnormal Sensation
2. Comments for Abnormal Sensation:

Comments (30)
I get stabbing pains around all areas affected.
By pressure I mean a sudden sensation as though something is pressing on the area. Not painful.
Ear sometimes feels like it's bleeding around the outside.
Strange sensations on my right forehead, top right of my head and lots of itching on right side of forehead and my right ear.
It can be annoying at times, sometimes my face will start twitching in areas, then I have this burning sensations and sometimes it feels as cold water was dropped on my face. The tingling and burning, I am having at this moment.
Affected area cannot be subjected to cold weather, will freeze much faster than normal skin.
The pain comes from the jaw difficulties I have. The tingling seems to be active in the back of my head above my ear. I can't be positive that the itching is related to PRS, but the affected area seems to itch more than other areas of my face.
At times the pain is so bad I feel like my head is being pried apart along the line of the indentation. The most pain killers can do at that point is take the hard edge off the pain. There doesn't seem to be a consistent "trigger" for when the pain will show up, except that it does appear to be linked to hormonal cycle, although not limited to these times. I think I am in some level of pain about 80% of the time, ranging from mild to severe. I could be having a completely normal day and out of nowhere the pain will start. It could stay mild and maybe it would be enough to go take a nap and wake up and it's gone, to coming on and becoming severe within minutes, to waking up in the night with severe pain. It's very unpredictable. Fatigue is also a predominate issue. I have to structure my day around making sure there is time to sleep during the day. Sometimes I require a couple extended naps a day. There is no choice in this, it's like a switch goes off and I have to sleep. If I force myself to stay awake, I usually end up with a headache, if I don't have one already.
I have those sensations when it's very cold or when i'm angry, when i have temperature, headaches or teeth pain.
Slight loss of sensation after surgery.
Right shoulder blade to waist affected - intense burning pain in shoulder blade area after repetitive activity.
If skin is bumped, then pain is much more than normal side of face.
Continued on the next page.

Page 17. III. 2. Comments (continued)
When active, I get a tingling in the affected area...top of head or nose.
I cannot sit with my head on my hand; then it hurts very much in my whole cranium - and I cannot lay very long on my pillow at night without pain.
I have tingling in my lip almost like an electrical zap. The burning is in my cheek area. It is not very strong or constant but it just feels odd.
I get severe headaces and pain around the eye, and also tingling in my right cheek and eye area along with twitches.
Loss of pain sensation in left leg - only just noticed it last week I need to get neurologist to check it out thoroughly, see if it affects the whole left side of my body (as the PRS is attacking the right side of my brain).
Diagnosed with hypotony of the left eye, very low pressure and DRY eye.
My face gets very itchy and just want to scratch it all the time. The tingling usually means that it is active and I can go for months without anything at all now.
Tingling...often associated with seizures.
My jaw tightens (like a spasm) and causes pain.
Periodic
Affected area is more sensitive to cold and to pain. There is pain and tingling (tingling even into the scalp) in the affected areas (particularly when it is activated).
I think that being in tune w/ your own body, you just know something is going on. I can't explain it, or maybe having had this for so long..I KNOW something is happening to me right now, but I cannot explain it- right now the right side of my forehead just over my eye has an odd sensation where there's been atrophy, and the left side of my face feels odd too where there's been atrophy. I just cannot explain it.
Extreme pain when lightly scratched or poked.
More like not-feeling the area.
Numbness and tingling but can't says thats it's only the PRS as I've had surgery to repair some of the damage.
In the previous question on the previous screen....I do not know if her blood pressure is affected. How could one know?
I am not sure what feelings she has or had.
I don't know if its from the Rombergs or not, but the skin on my face is very dry, especially in the cold months...............I also get an itchy rash on my face and neck from time to time...(feels like when one gets fiberglass on their skin)...sometimes my eyes will almost swell completely shut.......its been looked at by a dermatologist, but she couldn't figure out what it was or what was causing it.

Number responding = 30 / percent responding = 21% of total respondents (143).

Page 18. III. Associated Symptoms (page 1 of 4) cont.: Atrophy (tissue loss)

1. Please select the location of Atrophy (tissue loss) on the head:

Location	Responses (608)	% of Number responding
Cheek	105	81%
Chin	74	57%
Ear	27	21%
Eye lid	62	48%
Forehead	70	54%
Lower lip	37	28%
Upper lip	64	49%
Nose	56	43%
Scalp	57	44%
Temple	56	43%

Number responding = 130 / percent responding = 91% of total respondents (143).
Note: This was a multiple choice question.

Page 18. III. Associated Symptoms (page 1 of 4) cont.: Atrophy (tissue loss)

2. Other locations of the body with Atrophy (tissue loss):

Location	Responses (88)	% of Number responding
Arm	13	29%
Back	15	33%
Chest	9	20%
Leg	12	27%
Neck	16	36%
Side	5	11%
Stomach	4	9%
Other (Specify)	14	31%

Number responding = 45 / percent responding = 31% of total respondents (143).
Note: This was a multiple choice question.

Page 18. III. Associated Symptoms (page 1 of 4) cont.: Atrophy (tissue loss)
2. Other locations of the body with Atrophy (tissue loss):
Other (Specify)

% of Total Responses	Responses (14)	Other - Abnormal Sensation
2%	2	foot
1%	1	gum
5%	4	none
1%	1	not diagnosed on the body
1%	1	not sure
1%	1	ON RIGHT EYEBROW
1%	1	palate
1%	1	right breast
2%	2	tongue

Page 18. III. Associated Symptoms (page 1 of 4) cont.: Atrophy (tissue loss)
3. Please rate the severity of this Atrophy (tissue loss):

Rating	Responses (128)	% of Number responding
1 (very mild)	3	2%
2 (mild)	28	22%
3 (moderate)	57	45%
4 (severe)	30	23%
5 (very severe)	10	8%

Number responding = 128 / percent responding = 90% of total respondents (143).

Page 18. III. Associated Symptoms (page 1 of 4) cont.: Atrophy (tissue loss)

4. Comments for Atrophy (tissue loss):

Comments (48)
It is moderate in my leg but severe in my arm and face.
Body atrophy may be that the body never developed properly. Body atrophy is severe. Facial atrophy is mild.
First aware of atrophy at the age of 19 in my right breast.
Cheek followed skeletal line of face (no tissue left).
I would like to get something done to my face (filler, etc.), but I think that the disease is still active.
THE LOSS IS SO SIGNIFICANT THAT THE RIGHT EYE IS HIGHER ON MY FACE THAN THE LEFT AND IT HAS THE APPEARANCE OF BEING BIGGER AND "BUGGING OUT."
From age 11 onward, atrophy became much more noticable and tissue loss very significant- especially from mid cheek area on left hand side.
The whole side of my left face is sunk in because there is no fatty tissue there and it looks bluish at times.
What can we say it happens and there is no stopping it. You just have to go through with it. The symptons do stop for a while before they start back up again.
Continued atrophy of the chin.
Can't be positive that I have tissue loss of the leg and back due to PRS, but it seems odd that I have muscle problems on the left side of my body in these areas.
It's only skin and bone under my eye and you can feel the articulation under my chin and I have like small balls in the flesh that are hard but not all the time.
I feel that mine is still progressing. I can almost see a slight change in my forehead and cheek weekly.
The atrophy started with just tissue loss but then moved to bone loss in lower right jaw and tooth loss on lower right side.
Don't know what is considered mild or severe, so I said moderate.
Skin on back area is very thin & you can see the veins through it. Very little tissues under cheek area.
It seems the atrophy progressed rapidly as son approached puberty. I know that PRS is less common in males and therefore don't know if this experience w/puberty is common among other male patients. From my reading of other's emails, it seems that many females affected by PRS discuss increase in symptoms during pregnancy and/or menopause. Seems to be a hormonal link.
The top of my head is moderate to severe, but nobody sees that. My face is mild.
Since the tissue loss has just started, I would say it is moderate, but progressing rapidly.
Atrophy of entire left side of head including bone.
They say I have no tissue on effected side, and almost no bone mass there. Also I have little motor skills on that side of my face.
My atrophy is a lot worse now than it was when I was first diagnosed. It has spread further towards the back of my head and is also worse in the forehead.
Severe atrophy on the cheek.
On forehead, it is en coupe de sabre.
I still today get very upset when someone makes a comment about my face, people can be very cruel.
I feel it is severe, but compared to others I have seen, probably moderate.
Atrophy loss means that there is also skin discoloration which is very difficult to conceal or correct. Eye muscle loss leads to double vision and eye drooping lids and other eye complications that are not easily resolved.
Severe atrophy of forehead and scalp.
Loss of tissue is mainly around the chin and cheek area.

Continued on the next page.

Page 18. III. 4. Comments (continued)
Moderate to severe in some areas (under the affected eye).
Very severe at the injury site at the top of my forehead, the loss is down to the bone. In the rest of my face it is only moderate to very mild with tissue loss and it isn't noticeable until it is pointed out.
The places where I have less fat (e.g. my neck) are harder to see the atrophy.
Due to surgery hard to say the severity of atrophy.
I have had two fat injections into the right side of my face.....side of face, cheek, and chin. First set of injections in June 2008 in hospital under general anesthesia. Quite a bit of fat was reabsorbed. Second set in January during in-office procedure.
The atrophy makes it much more painful, if the affected side of my face is ever hit by anything (accidentally!). It also feels the cold much more.
Am not sure if I'd be mild or moderate. The odd thing is, given that they know so little about this, it really doesn't provide relief when they say they think it's mild b/c it keeps on going and no one knows how long it will go or how far it will go. It would help to know if not-mild cases generally have aggressive onset or if they all start out mild and then progress to greater stages.
Other areas may be affected but to a much lesser degree. I've only mentioned the moderately affected areas. Mouth has also been affected; roof of mouth is deeper on the left, tongue is thinner on left, gums receding on left.
My plastic surgeon, Dr. Mitchell Stotland of Dartmouth Hitchcock Medical Center in Lebanon, NH says that my atrophy is mild to moderate and that he's seen worse. Looking at a picture of myself that I took last night, that is hard for me to believe. I'm sure that he knows what he is talking about. I'll have to post the picture on the Romberg's Connection site as an updated one.
I would say it is moderate to severe. Has lost all tissue in his cheek. As his mother, I think it is severe....doctors would probably say moderate.
My eye has sunk and the area under the right eye is darker. The area in front of the ear was noticed first and the other side of my face was scanned for a tumour, because it was much plumper, particularly around the sinus area. However Dr Landy in Brisbane, an elderly neurologist, had come across hemifacial atrophy before and quickly diagnosed it. I have not had an MRI in seven years, so I do not know if the brain has atrophy. There were strange white spots on my MRI though.
Affected area is darker compared to the non affected area.
My daughter was in a researh program at New York University.
I have fillings with permanent material in my cheek but I think I have no natural fillings (fat tissue) left. Its just skin over bone on my jaw joint.
I had a face lift when I was 40 years old.
Loss of roots of teeth.
I also had tissue loss around the eye socket.
It doesn't bother me that much and I've seen much worse on some of the pictures from the group.........but the CAT scan of my head shows hardly any fat at all on the left side of my face. I have a narrow face anyway, so i guess it doesn't stand out as much....I have like a pit, in my cheek and ruts and grooves in my chin.
Severe loss, resulting in facial disfigurement, until surgery, which restored the tissue, but eventually reasorbed and back to moderate atrophy.

Number responding = 48 / percent responding = 34% of total respondents (143).

Page 19. III. Associated Symptoms (page 1 of 4) cont.: Blood pressure

1. Blood pressure is:

Pressure	Responses (11)	% of Number responding
High	9	89%
Low	2	11%

Number responding = 11 / percent responding = 8% of the total respondents (143).

Page 19. III. Associated Symptoms (page 1 of 4) cont.: Blood pressure

2. Other:

Comments (6)
optic neuritis, occulomotor problems
on the low side of normal.
asthma, allergies
Not sure if it's related to PRS.
Diagnosed with diabetes Aug. 07, so I was put on blood pressure meds.
Blood pressure was high ages 21-27 but has stabilized this past year.

Number responding = 6 / percent responding = 0.4% of total respondents (143).

Page 20. III. Associated Symptoms (page 1 of 4) cont.: Bone loss
1. Please select any location of bone loss:

Location	Responses (147)	% of Number responding
Cheek bone	36	55%
Forehead	24	36%
Jaw bone	41	62%
Temple	18	27%
Skull	28	42%

Number responding = 66 / percent responding = 46% of total respondents (143).
Note: This was a multiple choice question.

Page 20. III. Associated Symptoms (page 1 of 4) cont.: Bone loss
2. Other locations of bone loss:

Other locations	Responses (5)	% of Number responding
Arm	1	20%
Back	0	0%
Chest	0	0%
Foot	0	0%
Hand	1	20%
Hip	0	0%
Leg	0	0%
Ribs	0	0%
Spine	1	20%
Other (Specify) **None** *	2	40%

Number responding = 5 / percent responding = 0.3% of total respondents (143).
Note: This was a multiple choice question.
***** Two responses were "**None**" for "Other (Specify)".

Page 20. III. Associated Symptoms (page 1 of 4) cont.: Bone loss

3. Comments for Bone loss:

Comments (19)
Also chin of the affected side.
Saw a dentist last year who took x-rays. He said that there is substantial bone loss on lower right jaw, causing damage to the roots of the teeth in that area. He said that I will need to have a bone graft and dental implants done to replace the existing bridge which is at risk of breaking.
The plastic surgeon who diagnosed said the atrophy was in the left cheek and the jaw, but I don't notice this to be so. Must be very mild if it is.
Cheek bone did not develop as much as other cheek.
My worst atrophy is on the top and side of my skull.
The 3D cat scan showed the bone loss and the dental x-rays.
Appears like a 'sinking' in the area.
The majority of my bone loss is on my forehead.
Very slight
She needs bone implants along with teeth implants on left side top and bottom.
I was only visually diagnosed at this time.
Roughly 5cm diameter circle on top on head, left side.
Indentation about the size of an adult palm on skull on affected side. No hair loss on head.
Haven't been checked properly; only listed the clearly affected areas.
Replaced by plastic cheek bone. Probable slight bone loss all over right side of face also.
Palate inside the mouth atrophied... lost all top teeth.
Roots of two teeth on lower left jaw were resorbed. Wisdom tooth on lower left jaw was also abnormally small.
Bad teeth
Bone loss didn't occur until after brain surgery for seizures which resulted in speech loss, weaknes on right side and some bone loss on left side eventually.

Number responding = 19 / percent responding = 13% of total respondents (143).

Page 21. III. Associated Symptoms (page 1 of 4) cont.: Brain affected

1. Please select how brain is affected:

Affected	Responses (48)	% of Number responding
Atrophy of	7	26%
Lesions	8	30%
Seizures	12	44%
Strokes (blood clots)	2	7%
White matter abnormality	8	30%
Other (Specify)	11	41%

Number responding = 27 / percent responding = 19% of total respondents (143).
Note: This was a multiple choice question.

Note: If "Seizures" was selected at the above question 21. III. 1. "Please select how brain is affected" then the following section (Page 22) would apply.

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures

1. Please select the type/types of Seizures:

Type	Responses (25)	% of Number responding
Grand mal	8	62%
Jacksonian epilepsy (sensory seizures)	2	15%
Loss of feeling	3	23%
Petit mal	6	46%
Tingling sensations	4	31%
Other (Specify)	2	15%

Number responding = 13 / percent responding = 9% of total respondents (143).
Note: This was a multiple choice question.

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures
1. Please select the type/types of Seizures:
Other (Specify)

% of Total Responses	Responses (2)	Other - Type/Types of Seizures
4%	1	sharp pains, also get the shakes
4%	1	simple and some pseudo

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures
2. Please list any medications taken for seizures:

Comments (11)
TRILEPTAL 600MG DAILY, DOWN FROM 1200 MG
Dilantin (400 mg daily), but began having visual auras again after three years of no symptons). Keppra for a few weeks, slowly ramping up. Now Lamictal (400 mg daily) and no auras since November.
Phenobarbital at age 12 for about a month or so.
Keppra for migraines and seizures.
AUGMENTINE DEPAKINE (solución) DEPAKINE CRONO KEPRA LABILENO LUMINAL MYSOLINE NOOTROPIL NORMOVITE SABRILEX SIBELIUM SOMAZINA TEGRETOL TOPAMAX TRILEPTAL ZONEGRAN
Keppra Trilepta ** We are VERY careful NOT to use a generic brand which is NOT the same. Ativan for anxiety related to seizures. Clonazepam as a sleeping aid.
Currently taking Neurontin, Lamictal and Keppra. Have taken almost all anti-seizure medications in the past at one time or another.
100 mg. Topomax 2X a day. Once in the a.m., once in the evening before bed.
Tegretol (carbamazapin) 200 mg 3x a day Lamictal (lamotrigine) 100 mg 5x a day
Every kind, I can't list them all.
Keppra

Number responding = 11 / percent responding = 8% of total respondents (143).
Note: These medications with these four questions are combined in a chart below.

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures
3. Length of time medications were taken:

Comments (11)
11 YEARS
Dilantin, three years; Keppra for a few weeks; Lamictal 4 months
About a month
Over 1 year
13 years, taking 2 or 3 medication at a time.
6 months
Been on anti-seizure medicines since age 3.
Have gradually increased since October '08.
Will be taking them forever.
3 years
18 months

Number responding = 11 / percent responding = 8% of total respondents (143).
Note: These medications with these four questions are combined in a chart below.

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures
4. Please explain if these medications were or were not helpful:

Comments (11)
WERE HELFUL
Yes, they were and are very helpful. The Keppra, however, brought on severe psychotic reactions (severe hostility, depression and thoughts of suicide: all three of these were rare reactions which the paperwork cautioned users to contact the doctor immediately).
I didn't take them long enough to find out.
Keppra but they keep upping it because she needs to have less migraines. She has only had two seizures.
Probably Sabrilex was the best medication, that got for huge time Tasio free of seizures. It is difficult to say, how good some of the meds were, because I can't compare with not having taken meds at all.
Having breakthrough seizures...presently adjusting medication.
Seizures are intractable, but they would be even worse without medications.
So far, so good!
I am seizure free with this medication.
Not really, see previous answers. She had some seizure activity until the day she died.
Somewhat

Number responding = 11 / percent responding = 8% of total respondents (143).
Note: These medications with these four questions are combined in a chart below.

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures

5. What were some of the side effects of these medications:

Comments (11)
NONE THAT I REALIZED
Dilantin: swollen gums, mild fever (about 1 1/2 degrees) Keppra: severe psychotic reactions (see above) Lamictal: none so far
They made me sleep all the time.
You cannot miss a dose that can cause a seizure, have to take with food. Can cause I believe kidney problems.
somnolencia, irritabilidad, lentitud, torpeza .... (Note: A rough translation has been provided: "drowsiness, irritability, slowness, stupidity")
Tired
No response
Tingling in fingers and toes, but they made me lose weight, so I don't care!!!
Sleepiness, fatigue, forgetfulness, some loss of coordination at the beginning. The forgetfulness persists, other symptoms have faded.
Memory loss Kidney infections Immune system compromised Acting stoned Inappropriate behavior Slurred speech
No response

Number responding = 11 / percent responding = 8% of total respondents (143).
Note: These medications with these four questions are combined in a chart below.

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures

Combination of the four questions for these medications:

Medication	Length time taken	Helpful or Not	Side effects
TRILEPEPTAL 600MG DAILY, DOWN FROM 1200	11 YEARS	WERE HELPFUL	NONE THAT I REALIZED
Every kind, I can't list them all.	3 years	Not really, see previous answers. She had some seizure activity until the day she died.	memory loss kidney infections immune system commpromised acting stoned inappropriate behavior slurred speech
AUGMENTINE DEPAKINE (solución) DEPAKINE CRONO KEPRA LABILENO LUMINAL MYSOLINE Nootropil NORMOVITE SABRILEX Sibelium Somazina TEGRETOL TOPAMAX TRILEPTAL ZONEGRAN	13 years taking 2 or 3 medication at a time.	Probably Sabrilex was the best medication, that got for huge time Tasio free of seizures. It is difficult to say, how good some of the med where, because I can't compare with not having taken med at all.	somnolencia, irritabilidad, lentitud, torpeza .... (Note: A rough translation has been provided: "drowsiness, irritability, slowness, stupidity")
100 mg. Topomax 2X a day. Once in the a.m., once in the evening before bed	Have gradually increased since October '08	So far, so good!	Tingling in fingers and toes, but they made me lose weight, so I don't care!!! :)
Phenobarbital at age 12 for about a month or so.	about a month	I didn't take them long enough to find out.	They made me sleep all the time.
Keppra	18 months	Somewhat	No response
Keppra Trileptal ** We are VERY careful NOT to use a generic brand which is NOT the same Ativan for anxiety related to seizures Clonazepam as a sleeping aide	6 months.	Having breakthrough seizures...presently adjusting medication.	Tired
Currently taking Neurontin, Lamictal and Keppra. Have taken almost all anti-seizure medications in the past at one time or another.	Been on anti-seizure medicines since age 3.	Seizures are intractable, but they would be even worse without medications.	No response

Continued on the next page.

Medication	Length time taken	Helpful or Not	Side effects
Keppra for migraines and seizures	over 1 year	Keppra but they keep upping it because she needs to have less migraines. She has only had 2 seizures.	You cannot miss a dose that can cause a seizure, have to take with food. Can cause I believe kidney problems.
Dilantin (400 mg daily), but began having visual auras again after three years of no symptons). Keppra for a few weeks, slowly ramping up. Now Lamictal (400 mg daily) and no auras since November.	Dilantin, three years; Keppra for a few weeks; Lamictal four months.	Yes, they were and are very helpful. The Keppra, however, brought on severe psychotic reactions (severe hostility, depression and thoughts of suicide: all three of these were rare reactions which the paperwork cautioned users to contact the doctor immediately).	Dilantin: swollen gums, mild fever (about 1 1/2 degrees) Keppra: severe psychotic reactions (see above) Lamictal: none so far
Tegretol (carbamazapin) 200 mg 3x a day Lamictal (lamotrigine) 100 mg 5x a day	Will be taking them forever.	I am seizure free with this medication.	Sleepiness, fatigue, forgetfulness, some loss of coordination at the beginning. The forgetfulness persists, other symptoms have faded.

Number responding = 11 / percent responding = 8% of total respondents (143).

Page 22. III. Associated Symptoms (page 1 of 4) cont.: Seizures
6. Comments for Seizures:

Comments (7)
They happened only when I was sleeping, about every other month. My parents seemed to think that they would happen when I was overly tired or stressed.
I am always told they do not know if they are PRS related, same with migraines. How will I know if they are or they are not.
I think he has had all forms of seizures, from very little one to grand mal, from only one side, to big convulsions for a lot of time. All time the seizures affects his capacity of speaking and his right arm and hand.
Still struggling to find the right drug cocktail that will work.
I don't remember the seizure, so that didn't bother me- unfortunately it happened in front of my children though which wasn't good. The part that really has been hard was the loss of my driving privelege for 6 months, and not being able to work.
Grand Mal seizures out of the blue, in my sleep at 44.
All kinds, including the psuedo, eventually her body reacted to any problem particulalry fevers with seizure activity.

Number responding = 7 / percent responding = 5% of total respondents (143).

Page 21. III. Associated Symptoms (page 1 of 4) cont.: Brain affected
1. Please select how brain is affected:
Other (Specify)

% of Total Responses	Responses (11)	Other - How Brain is Affected
2%	1	cranial nerve damage
2%	1	small cyst
2%	1	STROKE BECAUSE OF DAMAGED BLOOD VESSELS
2%	1	still trying to find out
2%	1	Migraines and she has several cysts on back part of brain but along with everything they don't know if its PRS related.
2%	1	headaches
2%	1	chronic migraines and auras
2%	1	severe migraine headaches
2%	1	not sure
2%	1	Brain is smaller on affected side.
2%	1	Enlarged/inflamed cochlear nerve

Page 21. III. Associated Symptoms (page 1 of 4) cont.: Brain affected
2. Please describe what if any effective treatment was obtained:

Comments (18)
Avastin injections into the eye seemed to stop progression of optic nerve damage, but it did not cure the damage done.
None
I was prescribed ADHD (Attention-Deficit / Hyperactivity Disorder) medication (Ritalin) to help with brain function. I was told I have Organic ADHD because of Parry Rombergs Syndrome.
None
None
Lesion removed surgically, rest of smaller lesions reduced or eliminated by steroids (30mg every other day of Prednisone, infusion of 1000 mg of Methylprednisolone once per month). Methotrexate usage (20mg. once per week) to suppress over-active immune system.
I was not treated for the seizures since we were living in Jamaica at the time. When we returned to the US, the doctors put me on Phenobarbital, but my mother took me off of it since it made me sleep all the time and my last seizure was in Jamaica.
She is on Keppra, Methotrexate, folic acid, vitamin b6, Prilosec, Zantac, and just recently was taken off Prednisolone but may be put back on it and also she takes Naproxen.
I have been treated for chronic migraines and auras which my neurologist believes are related to Parry-Romberg.
For some time anti epileptic medicine were effective and he got no or only few seizures, but for long period - now since 3 years the anti epileptic medicines are no more effective, and he has gotten all kind of meds.
Seizure medication Pain medication
Pain relief
Inflammation in eye and face as well as appearance on her brain MRI was reduced with steroid therapy, both oral and IV pulse steroid.
After first, and thankfully only (so far) grand mal seizure, am taking 200 mg. of Topomax which is working well. This is being taken along w/ an 81 mg. aspirin, and meds for my Lupus. So far, so good!
Liquid silicone injections
Not sure if related to PRS or not, but cochlear nerve on affected side is enlarged.
Methotrexate - don't know if effective though can't imagine worse outcome than what we got.
Katie was on at least 8 different anti seizure meds plus steroid treatment and surgery, none of which worked, they would seem to work for awhile, but it was always short lived. Also increased seizure activity definitely around her cycle. Several times they put her into an anesthesia induced coma, to stop the seizures, but this ended up with her being on a respirator and finally trached, resulting in reccuring pneumonia.

Number responding = 18 / percent responding = 13% of total respondents (143).

Page 21. III. Associated Symptoms (page 1 of 4) cont.: Brain affected
3. Comments for Brain affected:

Comments (17)
Dizziness, vertigo, confusion, light-headed-ness, nausea
Left side of my brain is atrophied. I also have small lesions on brain. This was found on a MRI. Also at one point I had small cysts on my brain but they went away.
I haven't had a brain scan done, so don't know if the atrophy went into the brain.
Multiple lesions that seem to come and go, and one that's spread like little tentacles through portions of the right side and hasn't gone away entirely.
I had grand mal seizures. They took place only when I was sleeping. It would usually happen once about every other month. This lasted for about 3 years.
Still in proccess of being tested.
Typical PRS findings on MRI, whole right half of brain is smaller than left side. Particularly cerebellum and I am losing Motor Neurone on the PRS side causing spastic paralysis on the left side of my body. Also brain activity higher than normal - a sign that I could be prone to epilepsy although I've never had a seizure to date.
Severe migraines, seizures and she also gets these times where she just stares into space and drools.
She started getting alot of headaches when this first started, they have tapered off, but then she just had two this last week again.
Latest seizure activity difficult to locate on imaging.
Brain shows areas of calcification.
After IV pulse steroid therapy, progression in brain abnormalities were stopped and reversed slightly. Left temporal lobectomy was also done in effort to control intractable seizures.
Severe headaches since around 2004 - Doctors didn't know what caused them, and prescribed Indomethacin 50 mg. as needed which works pretty well. An MRI showed that I have three lesions back in '04 or '05, and after this seizure in Oct. '08, another showed that the lesions had changed a bit. Of course, they had to do tests for MS, etc. And, they are currently trying to figure out if my seizure was caused by the PRS, or by the Lupus... my guess is the PRS... Doctors only have me as a PRS patient- probably like all of the rest of you guys, huh?!
Not sure
Grand Mal Seizurs started in my 40's, happened only during sleep.
Negative neuro exam but does have changes on two consecutive MRI's.
Speech and memory loss, more so after the surgery to remove lesions which just made her worse.

Number responding = 17 / percent responding = 12% of total respondents (143).

Page 23. III. Associated Symptoms (page 1 of 4) cont.: Chin affected
1. Please select how the chin is affected:

Affected	Responses (180)	% of Number responding
Discoloration	30	41%
En coup de sabre (indentation)	47	64%
Thin skin	35	47%
Tissue loss	58	78%
Other (Specify)	10	14%

Number responding = 74 / percent responding = 52% of total respondents (143).
Note: This was a multiple choice question.

Page 23. III. Associated Symptoms (page 1 of 4) cont.: Chin affected
1. Please select how the chin is affected:
Other (Specify)

% of Total Responses	Responses (10)	Other - Chin Is Affected
1.7%	3	bone loss
0.6%	1	chin bone is off center
0.6%	1	crooked
0.6%	1	dark pigmentation
0.6%	1	fat
0.6%	1	lines that look like scars
0.6%	1	no hair, glowing
0.6%	1	puckering

Page 23. III. Associated Symptoms (page 1 of 4) cont.: Chin affected
2. Comments for Chin affected:

Comments (20)
One side seems to be smaller than the other.
Constant dull ache and extremely painful when banged.
I had a brown discoloration and it would turn blue in color when I was cold or exposed to water. I had a line or indention on left side of chin with tissue loss.
One part of my chin is bluish in color and it can look like it is a bruise. There is no fatty tissue on one part of it.
It was the second part affected and it has spread under it and goes on the neck. When the skin was hard, I had pains. When I tried to leave the hand on my back or simply just moving and problem swallowing food.
The plastic surgeon who diagnosed said it went through my chin, but I don't notice this. If that's true, it was very mild progression into the chin.
We originally thought my son "just had thin skin" on the right side of his chin. But as I mentioned before, it was as he approached puberty that the loss seemed to progress more rapidly and become more noticeable.
Chin is off center due to short mandible.
I have the en coup de sabre on my forehead.
I also get less "hormonal" pimples on the left side (PRS) of my chin (I get quite a lot on the right side).
I have had 3 fat injection treatments, but it seems that the chin is the hardest to correct. It does not seem to retain the fat very well, unlike a fleshier area like the cheek.
My skin on my chin has a slight indentation, but it's not hard. It started out as a small brown line and evolved into a depression that looks a bit pock-marky.
Chin cleft is severly off center.
The tissue around the chin had also atrophied.
Bone in the chin on one side was affected, gave me a slight indention.
Entire half of chin affected.
I had facial reconstruction when I was thirty years old.
Some people have asked if I have a crooked scar on my chin...............
There may be some bone loss...has not been diagnosed however.
These areas did not react to the silicone injections. It's painful if touched too hard.

Number responding = 20 / percent responding = 14% of total respondents (143).

Page 24. III. Associated Symptoms (page 1 of 4) cont.: Ears affected
1. Please select how the ear is affected:

Affected	Responses (49)	% of Number responding
Hearing loss	14	🟥 33%
Some atrophy of the ear canal	13	🟦 31%
Other (Specify)	22	🟥 52%

Number responding = 42 / percent responding = 29% of total respondents (143).
Note: This was a multiple choice question.

Page 24. III. Associated Symptoms (page 1 of 4) cont.: Ears affected
1. Please select how the ear is affected:
Other (Specify)

% of Total Responses	Responses (22)	Other - How the Ear is Affected
2%	1	slightly less cartilage
2%	1	Not sure on the ear: son had skin over eardrum get so tight it almost burst.
2%	1	protruding ear
2%	1	different size and shape
2%	1	Platform of ear is losing tissue.
2%	1	cartilage loss
2%	1	missing ear lobe
2%	1	outer ear tissue loss only
2%	1	external tragus and ear lobe
4%	2	atrophy of the outer ear
2%	1	off center
2%	1	fuzzy numbness in ear
4%	2	hole in eardrum
4%	2	Ear sticks out.
2%	1	not affected
6%	3	thin ear
2%	1	droopy

Page 24. III. Associated Symptoms (page 1 of 4) cont.: Ears affected
2. Comments for Ears affected:

Comments (24)
One ear smaller than other.
During my 40's, my ears were tested. The result was a scar on the eardrum of a hole that healed over time. I remember as a child I had sometimes severe earache, which was treated at home.
It's hard for me to hear sometimes, but I was tested and they told me I do not have hearing loss. When I was a child I had chronic ear infections and at one point I had a small hole in my ear due to ear infections. Pus would drain out of my ear canal and the brown spot formed on my neck all at age 8.
Surgery "T Tube" was used to alleviate tight skin over eardrum.
Right ear itself is smaller.
Suspect that parts of the sinus and inner ear are atrophied.
The ear always feels like it is blocked.
no effects
Tinnitis and Meniéres - and pain. The pain is rare - put very strong!
Lost hearing in left ear and needed a hearing aid in order to hear. After surgically moving both upper and lower jaw her hearing returned where she no longer needs the hearing aide. I don't know if the the bones were pressing on the nerve and when moved relieved the pressure on the nerve.
The cartilage is thinner.
The opposite side ear has an almost non-existent ear lobe.
Her hearing is slightly less in the left ear and she has never felt an ear infection in that ear but she does feel them in the right ear.
Low to moderate hearing loss in right ear. Uses FM Device in school.
My hearing isn't affected as far as I know.
Ear is smaller and ear canal is narrower.
Hearing loss is undiagnosed at this time, but I have noticed hearing loss over the last year or so (going back to the specialist later this month and will discuss).
Ear drum, there is scar tissue.
Left ear is smaller than the right.
atrophy
Everything is muffled... but can't be sure that the PRS caused this as I've had surgery and they cut RIGHT next the ear.
I got the hearing loss but it was due to acoustic neuroma, not Rombergs. The doctor said it's unlikely to associate with Rombergs because the tendency is opposite-- atrophy VS tumor.
One ear is thinner than the other and is positioned differently. Also almost no lower part of the ear (where the earring goes). Hearing is not affected.
Ear sticks out due to sinking in near mandible.

Number responding = 24 / percent responding = 17% of total respondents (143).

Page 25. III. Associated Symptoms of Rombergs (page 2 of 4)

1. Please select all that apply (page 2):

Symptoms	Responses (575)	% of Number responding
En coup de sabre (indentation)	92	71%
Eye affected	82	63%
Face affected	118	91%
Fasciculation (muscle twitching)	41	32%
Fatigue	42	32%
Forehead affected	75	58%
Hair affected	60	46%
Headaches	65	50%

Number responding = 130 / percent responding = 91% of total respondents (143).
Note: This was a multiple choice question.

Page 26. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 1 of 2)

1. Please select the location of En coup de sabre:

Location	Responses (159)	% of Number responding
Cheek	20	22%
Forehead	56	62%
Chin	33	37%
From scalp to chin, right side	9	10%
From scalp to chin, left side	13	14%
Tongue	14	16%
Other (Specify)	14	16%

Number responding = 90 / percent responding = 63% of total respondents (143).
Note: This was a multiple choice question.

Page 26. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 1 of 2)
1. Please select the location of En coup de sabre: Other (Specify)

% of Total Responses	Responses (14)	Other - Location of En coup de sabre
1.3%	2	forehead to crown
0.6%	1	forehead to top lip and back into my scalp
0.6%	1	from ear to chin
0.6%	1	from forehead into scalp
0.6%	1	from forehead to scalp on left side
0.6%	1	Have slight indent on forehead, but area is not hard, so don't know if that qualifies as en coup de sabre; some say it has to be hard tissue.
0.6%	1	head
1.3%	2	just scalp
0.6%	1	right head-opposite PRS side
0.6%	1	scalp, forehead, nose, teeth
0.6%	1	sunken eye
0.6%	1	upper lip nose

Page 26. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 1 of 2)
2. Where did it start:

% of Total Responses	Responses (83)	Where did it start:
1.2%	1	Around 1997
1.2%	1	Around lip and nose areas
1.2%	1	Around the eyes and drooping eyelid and whole of left side of face
1.2%	1	Blue discoloration
1.2%	1	Both face and back progression started in the same week after accidents.
1.2%	1	Center of chin
7.2%	6	Cheek
4.8%	4	Chin
1.2%	1	Chin / right side of face in general
1.2%	1	Chin and cheek
1.2%	1	Chin with a dark spot glowing spot
1.2%	1	Crown of head
1.2%	1	Discoloration under left eye and on stomach
1.2%	1	Discoloring of neck, hardness of skin on cheek

Continued on the next page.

% of Total Responses	Responses (83)	Where did it start:
1.2%	1	Don't know for sure. Looking at it, I would guess in the forehead.
1.2%	1	Droopy eye lid, white patch of hair
1.2%	1	Ear
1.2%	1	En coup de sabre was first noticed on left side of face.
1.2%	1	Eye and chin
1.2%	1	Eye brows
1.2%	1	Face/ cheek
12.%	10	Forehead
7.2%	6	Forehead-mid
1.2%	1	From between my eyes to the top of my forehead.
1.2%	1	Hair loss on the eyebrow and a white spot on the forehead above the eyebrow.
1.2%	1	I believe it started in the cheek and has now progressed to the temple.
1.2%	1	In her hair
1.2%	1	In my upper lip and eye
1.2%	1	Initially started at left upper lip
1.2%	1	Just below bottom lip
1.2%	1	Just noticed a ridge running down the left side of my face one day, and there wasn't one on the other side.
1.2%	1	Left arm
1.2%	1	Left chin area- near mouth
1.2%	1	Left upper lip
1.2%	1	lower cheek
1.2%	1	Mid forehead and proceeded into scalp with bone loss in jaw
1.2%	1	My cheek, followed by the eye
2.4%	2	Neck
3.6%	3	Not sure
1.2%	1	On my back
1.2%	1	On the chin and on the scalp
1.2%	1	On upper lip
1.2%	1	Possible chin
1.2%	1	Probably in the cheek or forehead - possibly with a discolored spot on my back which then atrophied
1.2%	1	Probably loss of hair
1.2%	1	Right side of chin
1.2%	1	Right side of lower back
2.4%	2	Scalp
1.2%	1	Scalp with a white spot and hair loss on left temple
1.2%	1	Small quarter size blemish on left jaw bone area
1.2%	1	Temple / cheek

Continued on the next page.

% of Total Responses	Responses (83)	Where did it start:
1.2%	1	The lower part of the left side of my lip in the corner
1.2%	1	Top of head (scalp)
1.2%	1	Under eye
1.2%	1	Under right eye, looked like a red rash
1.2%	1	Upper right side of forehead
1.2%	1	With a line of scleroderma on the cheek and ear

Number responding = 83 / percent responding = 58% of total respondents (143).

Page 26. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 1 of 2)

3. At what age:

Age	Responses (84)	% of Number responding
45 - 49	1	1%
40 - 44	0	0%
35 - 39	4	5%
30 - 34	1	1%
25 - 29	5	6%
20 - 24	5	6%
15 - 19	5	6%
10 - 14	19	23%
5 - 9	22	26%
1 - 4	17	20%
At birth	1	1%
Don't Know	4	5%

Number responding = 84 / percent responding = 59% of total respondents (143).

Page 26. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 1 of 2)

4. Where did it progress to:

% of Total Responses	Responses (79)	Where did it progress to:
1.3%	1	Around the chin up into the cheek and now into the forehead.
1.3%	1	Bottom of chin
1.3%	1	Cheek
1.3%	1	Cheek, ear, nose eyelid
1.3%	1	Cheek, forehead, chin, face, body
1.3%	1	Cheek, right nostril, recently forehead
1.3%	1	Cheek/scalp
5.1%	4	Chin
1.3%	1	Chin, eye
1.3%	1	Chin and tongue
1.3%	1	Chin, jaw, cheek
1.3%	1	Complete left chin up to near by the ear

Continued on the next page.

% of Total Responses	Responses (79)	Where did it progress to:
1.3%	1	Down and across right side of face and back in my scalp
1.3%	1	Down between her eyes
1.3%	1	Down forehead and around left ear and then left cheek
1.3%	1	Down into neck area, up around nose, around eye, cheek area, lips-right forehead- eyebrow
1.3%	1	Down to the bottom of my forehead
1.3%	1	Entire right side of face and under chin.
1.3%	1	Entire right side of face up into hair
1.3%	1	Entire side of face and spots of atrophy on body
1.3%	1	Everything!
1.3%	1	Eye
1.3%	1	Eye lid
1.3%	1	Eye, scalp, chin
1.3%	1	Face
1.3%	1	Face, neck, arm, chest wall, leg
3.8%	3	Forehead
1.3%	1	Forehead, chin
1.3%	1	Forehead, eye, chin, lips, ear
1.3%	1	Forehead, lips, nose, tongue, eye
1.3%	1	Forehead, then cheek
1.3%	1	From cheek to chin
1.3%	1	From the left eye, to the left side of the crown of my head, creating a big dent.
1.3%	1	Front scalp area to chin.
1.3%	1	Full side of the cheek, temple
1.3%	1	Further onto lip and into cheek and then upper forhead
1.3%	1	Head and chin
1.3%	1	Indentation on right side of scalp and loss of hair
1.3%	1	It progressed all over the left side of my skull.
1.3%	1	Left eye, left cheek then chin
1.3%	1	Left leg got a slight indent following pain, then chin showed change-pigment changes on face.
1.3%	1	Left side of face, eye, nose, forehead, scalp, left breast, back, stomach
1.3%	1	Length of face
1.3%	1	Lip, nose
1.3%	1	MOVE DOWN TOWARD THE BRIDGE OF MY NOSE AND UPWARD TOWARD MY SCALP. BY AGE 7, IT HAD INDENTED AND MOVED AN INCH OR SO INTO MY SCALP CAUSING ALOPECIA. MOVING DOWNWARD THE HAIR ON ONE SIDE OF MY EYEBROW IS GONE AND ALSO SOME OF MY EYELASHES. MY NOSE IS VERY ASYMETRICAL.
1.3%	1	More atrophy of the cheek and chin, then temple and forehead
1.3%	1	Mostly up and back into the skull
1.3%	1	My whole left side of face, arm, & leg

Continued on the next page.

% of Total Responses	Responses (79)	Where did it progress to:
1.3%	1	Nose
1.3%	1	Nose, inside mouth
1.3%	1	Nose, chin
1.3%	1	PRS symptoms
1.3%	1	Right cheek and then right upper thigh
2.5%	2	Right side of face
1.3%	1	Same
1.3%	1	Same spot
2.5%	2	Scalp
1.3%	1	Scalp to chin to forhead to tongue
1.3%	1	Stayed in area
1.3%	1	Stayed in scalp
1.3%	1	Still ongoing
1.3%	1	Teeth
1.3%	1	The side of face, in different areas on face
1.3%	1	The temple
1.3%	1	To cheek and forehead
1.3%	1	To scalp and through eye, along nose to chin
1.3%	1	To the back of my head and also to my nose
1.3%	1	Top of chin
1.3%	1	Top of skull
1.3%	1	Unknown
1.3%	1	Up to top of head
1.3%	1	Whole left side of face, cheek, side of nose temple

Number responding = 79 / percent responding = 55% of total respondents (143).

Page 26. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 1 of 2)
5. At what age did it appear to stop:

Age	Responses (83)	% of Number responding
45 - 49	3	4%
40 - 44	2	2%
35 - 39	1	1%
30 - 34	6	7%
25 - 29	8	10%
20 - 24	8	10%
15 - 19	8	10%
10 - 14	9	11%
5 - 9	3	4%
Not Sure	11	13%
Still Active	24	29%

Number responding = 83 / percent responding = 58% of total respondents (143).

Page 26. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 1 of 2)
6. Was there any hair loss before or after:

Hair loss	Responses (87)	% of Number responding
Before	10	11%
After	20	23%
Before and After	18	21%
None	39	45%

Number responding = 87 / percent responding = 61% of total respondents (143).

Page 27. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 2 of 2)
1. Was there any skin discoloration before or after:

Discoloration	Responses (87)	% of Number responding
Before	25	29%
After	15	17%
Before and After	24	28%
None	23	26%

Number responding = 87 / percent responding = 61% of total respondents (143).

Page 27. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 2 of 2)
2. Any medication or procedure that stopped the progression:

% of Total Responses	Responses (50)	Any medication or procedure that stopped the progression
2%	1	I WAS GIVEN ANABOLIC STEROIDS.
2%	1	I was not diagnosed until I was 23. The progression had stopped and no medication was given.
2%	1	I haven't got to be under a doctor for this just got to see a cosmatologist and he couldn't help me.
2%	1	We have tried Methotrexate but hard to judge the progession or abatement of PRS.
2%	1	Methotrexate and Prednisone oral tablets Dovonex cream
2%	1	On meds now to slow down progression. Do I think they work, no. Methotrexate, folic acid, Prednisolone, Naproxen, Prilosec, Zantac, Keppra, vitamin B6, Albutedol, I think that's all.
2%	1	Methylprednisolone and Methotrexate
2%	1	I haven't tried any medication specifically for PRS- taking too much as it is for Lupus already! Although, I'm currently on Plaquenil- isn't that a medication that is prescribed for PRS?? I have had a free- flap to face surgery in 1995 using a flap from my scapular area done by Dr. Julian Pribazz of Brigham and Women's Hospital in Boston, MA. I've also had several fat injection surgeries by Dr. Stotland, and he excised my "bald spot" on the en coup de sabre and tried to sew up the "bald spot" in my right eyebrow, but now that's gone. He also rebuilt my left eye socket as it had atrophied, and I had a procedure during which he and another Dr. had moved my left eye up and took a piece of cartilage from my right ear and used it in my lower left eye lid to bring it out, etc. I'm really just a piece of work!!
2%	1	Removal of initial affected area on lip and gum, they left area on nose etc.
2%	1	Methotrexate & Predisolone, seems like it stops off & on.
2%	1	Not sure anything helped.
2%	1	High doses of Penicillin for two weeks i. v. Suppose that it stopped for about 5 years, then started again. Process of progression is very slowly.
2%	1	Steroids were tried when I was first diagnosed, but appeared to have no effect. I have had 3 fat injections, which do not stop the progression but certainly help the aesthetics.

Continued on the next page.

% of Total Responses	Responses (50)	Any medication or procedure that stopped the progression
2%	1	Steroids
2%	1	A cream was applied to the area where it started to burn it down. It was raised and after using the cream, it is now smooth.
2%	1	Yes Methotrexate
2%	1	Microvascular free flap transfer
2%	1	She takes Methotrexate and folic acid, and was on Prednisolone. It has slowed down but did not stop that is what doctors say. I honestly have not noticed it stopping sometimes I think it has gotten worse.
2%	1	As far as I can recall I was put onto Diprosone (it was a medication given to leppers).
2%	1	None, I have been told there is nothing to stop the progression.
2%	1	Methotrexate tablets and IV Methylprednisolone
2%	1	Was on Doxy on and off for lyme. During that, changes appeared to arrest. Stopped looking, although fasciculations continued body-wide. Was told they were benign fasciculations. Then went thru stressful period (still in it) and noticed chin changes. Had been off Doxy for a year by then.
2%	1	I take extra vitamin D, which I think helps. Also an aspirin per day for the vascular dilation to the area.
54%	27	None

Number responding = 50 / percent responding = 35% of total respondents (143).

Page 27. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 2 of 2)

3. Please comment on effectiveness of this medication or procedure.

% of Number responding	Responses (27)	Effectiveness of Medication or Procedure
3.7%	1	A picture is worth 1000 words. I'll post one for you to decide. I think that the free flap has held the best so far, hard saying about the rest- seems as if the PRS just keeps on taking back what has been done. That, or i'm just going to either keep on having surgery over and over, or just have to like what I look like no matter what it is.
3.7%	1	During this time I had weekly blood counts. Whether it was this medication that halted the atrophy or whether it just did not progress any further one would never know.
3.7%	1	Ear
3.7%	1	First dose of oral steroids had a dramatic effect on eye inflammation. Ptosis was gone immediately, and redness of conjuctiva disappeared. Any time we have tried to wean her off the small maintenance dose of oral steroids, inflammation returns.
3.7%	1	Hard to say for sure.
3.7%	1	I don't know what to think.
3.7%	1	I WAS GIVEN ANABOLIC STEROIDS. THEY CAUSED THEIR OWN PROBLEMS
3.7%	1	It seemed to stop advancement nose continued to regress but the area affected didn't continue to grow.
3.7%	1	It seemed to stop or at least slow the progression.
3.7%	1	It was very effective in halting the disease and in the reconstruction of my face.
18.5%	5	N/A
3.7%	1	No medication or procedure was effective in treating Parry-Romberg.
3.7%	1	No meds
3.7%	1	Not effective in my opinion.
3.7%	1	Not helpful
3.7%	1	See above
3.7%	1	Seems to be inhibiting further progression.
3.7%	1	She is currently taking Prednisone and Methotrexate for treatment.
3.7%	1	Sometimes I wish I would have never put her through all these medications. She gaines a lot of weight with the Prednisolone and now has asthma, sinus problems, her heart races. She didn't have these problems before.
3.7%	1	The lines of Scleroderma are softer and brighter, the regression of half face progresses.
3.7%	1	The liquid silicon injections helped fill in partial areas of face, but have left her with problems with infections and spaces between tissue and treated areas.
3.7%	1	The steroids were ineffective.
3.7%	1	Told me to use a topical cream, but it didn't stop it.
3.7%	1	Very effective

Number responding = 27 / percent responding = 19% of total respondents (143).

Page 27. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 2 of 2)

Combine the "Medication or Procedure" (Page 27.2) response with the "Effectiveness" (Page 27.3) response.

Medication or Procedure (Page 27.2)	Effectiveness of Medication or Procedure (Page 27.3)
NO	I WAS GIVEN ANABOLIC STEROIDS. THEY CAUSED THEIR OWN PROBLEMS
I haven't got to be under a doctor for this just got to see a cosmatologist and he couldn't help me.	Told me to use a topical cream, but it didn't stop it.
Methylprednisolon and Methotrexate	T he lines of Scleroderma are softer and brighter, the regression of half face progresses.
None	None
I take extra vitamin D, which I think helps. Also an aspirin per day for the vascular dilation to the area.	Hard to say for sure.
None	n/a
Microvascular free flap transfer	It was very effective in halting the disease and in the reconstruction of my face.
She takes Methotrexate and folic acid, and was on Prednisolone. It has slowed down but did not stop that is what doctors say. I honestly have not noticed it stopping sometimes I think it has gotten worse.	Sometimes I wish I would have never put her through all these medications. She gaines a lot of weight with the Prednisolone and now has asthma, sinus problems, her heart races. She didn't have these problems before.
Removal of initial affected area on lip and gum, they left area on nose etc.	It seemed to stop advancement, nose continued to regress but the area affected didn't continue to grow.
None	No medication or procedure was effective in treating Parry-Romberg.
Methotrexate and Prednisone oral tablets Dovonex cream	Seems to be inhibiting further progression.
Yes Methartrexate	Very effective
Steroids were tried when I was first diagnosed, but appeared to have no effect. I have had 3 fat injections, which do not stop the progression but certainly help the aesthetics.	The steroids were ineffective.
Steroids	First dose of oral steroids had a dramatic effect on eye inflammation. Ptosis was gone immediately, and redness of conjuctiva disappeared. Any time we have tried to wean her off the small maintenance dose of oral steroids, inflammation returns.
Was on Doxy on and off for Lyme disease. During that, changes appeared to arrest. Stopped looking, although fasciculations continued body-wide. Was told they were benign fasiculations. Then went thru stressful period (still in it) and noticed chin changes. Had been off Doxy for a year by then.	I don't know what to think.

Continued on the next page.

Medication or Procedure (Page 27.2)	Effectiveness of Medication or Procedure (Page 27.3)
No	N/A
We have tried Methotrexate but hard to judge the progession or abatement of PRS.	see above
Methotrexate tablets and IV Methylprednisalone	It seemed to stop or at least slow the progression.
"I haven't tried any medication specifically for PRS- taking too much as it is for Lupus already! Although, I'm currently on Plaquenil- isn't that a medication that is prescribed for PRS?? I have had a free- flap to face surgery in 1995 using a flap from my scapular area done by Dr. Julian Pribazz of Brigham and Women's Hospital in Boston, MA. I've also had several fat injection surgeries by Dr. Stotland, and he excised my "bald spot" on the En coup de sabre and tried to sew up the "bald spot" in my right eyebrow, but now that's gone. He also rebuilt my left eye socket as it had atrophied, and I had a procedure during which he and another Dr. had moved my left eye up and took a piece of cartilage from my right ear and used it in my lower left eye lid to bring it out, etc. I'm really just a piece of work!!"	A picture is worth 1000 words. I'll post one for you to decide. I think that the free flap has held the best so far, hard saying about the rest- seems as if the PRS just keeps on taking back what has been done. That, or i'm just going to either keep on having surgery over and over, or just have to like what I look like no matter what it is.
On meds now to slow down progression. Do I think they work, no. Methotrexate, folic acid, Prednisolone, Naproxen, Prilosec, Zantac, Keppra, vitamin B-6, Albutedol, I think that's all.	Not effective in my opinion.
No	N/A
Not sure anything helped	The liquid silicon injections helped fill in partial areas of face, but have left her with problems with infections and spaces between tissue and treated areas.
"I don't think I have any discoloration other than the red chapped skin.......not sure if that's associated with it or not.......No meds."	No meds
As far as I can recall I was put onto Diprosone (it was a medication given to leppers).	During this time I had weekly blood counts. Whether it was this medication that halted the atrophy or whether it just did not progress any further one would never know.
No	Not helpful

Number responding = 20 / percent responding = 14% of total respondents (143).
Note: Had to have answered both 27.2 and 27.3 to be included here.

Page 27. III. Associated Symptoms (page 2 of 4) cont.: En coup de sabre (page 2 of 2)

4. Please rate the severity of the En coup de sabre:

Rating	Responses (86)	% of Number responding
1 (very mild)	5	6%
2 (mild)	20	23%
3 (moderate)	38	44%
4 (severe)	19	22%
5 (very severe)	4	5%

Number responding = 86 / percent responding = 60% of total respondents (143).

5. Comments for En coup de sabre:

Comments (23)
The en coup de sabre start of indention was at the site where i got struck in the face. The hole in right side of my back also started at place I was struck. I still have a small white scar there.
IT IS A VIAL THING THAT HAS MADE ME SELF-CONSCIOUS ALL MY LIFE. I'VE NEVER BEEN ABLE TO PULL MY HAIR BACK BECAUSE OF THIS HUGE INDENTATION AND SCAR. ALSO, WHERE THE FAT AND BONE ARE AFFECTED IT IS VERY TENDER TO THE TOUCH.
To me it looks mild on my forehead.
It is like the dividing line between my Rombergs side and the normal side.
I have become very uncomfortable and self-conscious about my forehead now. I feel like it is drastic and wear my bangs down to cover it every day. I feel like it is "severe" but I have seen pictures of other people who were much worse... so I feel like severe would not be appropriate to write, compared to some others.
I say moderate because I don't know what is mild and what is severe.
It seems mild compared to others. I cover my forehead with hair (bangs). I have no masseter muscle on my left cheek. (MRI shows it missing).
Moderate on top of head, mild to moderate on forehead
Four streaks going down the front of my forehead
Crown of head is severe in flatness... forehead is very mild
Because of the severity of this, I make sure my hair covers my entire forehead and use hairspray to keep it in place.
This disease is very confusing.
From photos I have seen of en coup de sabre, her's seems to be pretty severe and quick spreading.
Stops right below eyebrow level.
From injury site mid-forehead down my forehead, nose and upper lip. About 1.5 cm wide.
My forehead and chin are quite noticeable.
It starts right on the top of my scalp and follows a line down my forehead.
I don't think I should judge the severity because I have nothing to compare to. Seems bad to me.
It was a big ol' dent in my head that used to really bug me because it was bald - I was very self conscious of it unit my Dr. excised it and stitched the skin together so that my hair covered it - now it isn't noticable.. When you push on the dent it causes pain though. Other than that, no problems.
Has a crooked chin at age 8, the doctors indicated PRS but nothing was done as only the chin was affected at the time.
She has very visible deterioration and has physical problems.
On the forehead and the chin.
Chin

Number responding = 23 / percent responding = 16% of total respondents (143).

Page 28. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 1 of 2)

1. Please select how is the eye affected:

Affected	Responses (298)	% of Number responding
Blurred vision	32	42%
Cataracts	10	13%
Corneal scarring	6	8%
Corneal transplant	0	0%
Detached retina	2	3%
Dilated pupil	16	21%
Discharge	10	13%
Double vision	14	18%
Drooping eyelids	34	45%
Glasses	36	47%
Heterochromia (difference in coloration, usually of the iris)	8	11%
Iridocyclitis (type of anterior uveitis)	2	3%
Iritis (inflamed iris)	5	7%
Keratitis (dry eye)	25	33%
Optic nerve damage	12	16%
Partial or complete loss of vision	14	18%
Receding eye	38	50%
Scleritis (primary inflammation of the sclera)	3	4%
Uveitis (inflammation)	7	9%
Other (Specify)	24	32%

Number responding = 76 / percent responding = 53% of total respondents (143).
Note: This was a multiple choice question.

Page 28. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 1 of 2)
1. Please select how is the eye affected:
Other (Specify)

% of Total Responses	Responses (24)	Other - How Is The Eye Affected
0.3%	1	A feeling of strain and irritation. When this is active I can only read a couple pages and then have to take a break and rest the affected eye or the feeling of strain can become severe. The eye becomes various levels of red depending on the amount of strain. Looking at the degree of redness in the eye is one indicator to me of the level of strain and the need to rest the eye.
0.3%	1	astigmatism
0.3%	1	born with squint
0.3%	1	Can't close lid all the way.
0.3%	1	Dr. says eye on affected side has much worse vision, and eye is different internally on that side as well.
0.3%	1	enopthalmus
0.3%	1	episcleritis
0.3%	1	eye movement
0.3%	1	eye twitches
0.3%	1	Eyelid and lashes are half gone.
0.3%	1	Her left eye has a tendancy to move outward and it also moves back and forth faster that the human eye can see.
0.3%	1	Lazy eye, not sure if optic nerve is affected.
0.3%	1	Left (affected) eye opening is larger, more "open" than the right.
0.3%	1	lens replacement
0.3%	1	Lower pressure in the left eye than the right eye. Usually half the normal pressure. Usually a pressure of 10 or less.
0.3%	1	No fat tissue in eyelids.
0.3%	1	No inner eye lashes, constantly itchy.
0.3%	1	not sure
0.3%	1	occulomotor problems
0.3%	1	Pupil in my affected eye is misshapened, almond shaped.
0.3%	1	retinal tears, no tearing
0.3%	1	Right eyeball is growing shorter than left one.
0.3%	1	strabismus
0.3%	1	Vitreous detachment in Romberg's side eye and flashing lights.

Page 28. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 1 of 2)

2. Please comment if the affected person had any surgeries that caused problems with their eyes:

Count	Comments (21)
1	A cream was applied to the area where it started to burn it down. It was raised and after using the cream, it is now smooth.
1	Cataract brought on the detached retina.
1	Droop was surgically corrected somewhat successfully.
1	I am sorry that I cannot answer this question - I know too little about the words.
1	I had my left eye socket rebuilt which caused my vision to be blurred, but it straightened out.
1	I have had cataract surgeries on both eyes. Only have PR on right side, which I believe started after the cataract surgery. I also had laser surgery to clear up blurriness.
1	I have had four surgeries to correct double vision on my left eye. One surgergy to correct a filtering bleb on the sclering. One surgery on the right eye in order to correct the double vision on the left eye. One surgery to correct the drooping lid on the upper left eye. One surgery to pick up the drooping lid on the lower left eye. I am about to have one final surgery to pick up the drooping lid on the upper left eye. Hopefully this is the end. But I have a cataract that will need surgery.
1	Just had lower lid tightened.
1	My eye surgeon, who performed my second surgery for my squint noticed that my left eye socket and eye ball were smaller (back when I was 26), but my doctor didn't follow up on it at the time. (I had my first surgery when I was 4, but the atrophy had not started at that time.)
1	My right eye was damaged in the first plastic surgery.
7	No surgeries
1	Not at this time but has vision problems.
1	The reconstructive surgery I had caused a lot of scarring around the outside of the eye... my tear duct was compromised, and the optic nerve was damaged.
1	When I got the filling material I could close my eye better. Over the years I got problems with dry eye, and shedding tears (seems contradictional).
1	Yes. Although the right eye always looked different, it never caused me pain until I had my first reconstructive surgery.

Number responding = 21 / percent responding = 15% of total respondents (143).

Page 28. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 1 of 2)
3. If the affected person had cataract surgery, please comment on the results:

Count	Comments (14)
1	I had a rare cataract and it took the surgeon a longer time to do and he had to use a more powerful machine than would with other patients. I could see a little more out of it until I had the detached retina.
1	I had PRK to help me with my vision. My left eye is lower then my right and so I had problems seeing out of glasses and contacts irritated my eyes. I was never able to see well and when i was pregnant it was worse. I just got PRK done and I am able to see for the first time in my life 20/16 in left eye and 20/18 in right eye.
1	I had small cataracts in both eyes and opted for two lens replacements. The left eye has been wonderful, but as I expected, I had problems with the right. It was done for long sight and and I have had laser surgery since to improve the sight, which it did, then it deteriorated quickly again, so I had a capsulotomy where a tiny slit is lasered in the back of the capsule to let the light through. This has worked well. My right eye is much drier than the left though and I have to lubricate it frequently. I am still getting pain through the right eye. I did not mention my Rombergs to the surgeon as I felt it would serve no purpose.
1	I have not had cataract removed yet in that eye.
8	No cataracts
1	See above
1	Yes, at age 5, no signs of PRS were present then.

Number responding = 14 / percent responding = 10% of total respondents (143).

Page 28. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 1 of 2)
4. If the affected person had any loss of vision, please comment:

Count	Comments (32)
1	Affected eye is weaker than good eye, but is my dominant eye.
1	Blurred vision, weeping of eye mostly when lying down.
1	Double vision now corrected after surgery. Small pupil (Horners Syndrome). Blurred vision in left eye
1	Has no peripheral vision on her left side.
1	I am almost blind in my right eye.
1	I had trouble getting fitted with the right lens prescription for my glasses the last time my eyes were examined by my Opthomologist. I had to have my eyes examined at least five different times. I would seem to be able to read the chart in the doctor's office, but after picking up my new glasses, had trouble seeing with them.
1	I have monocular vision and only use my left eye for peripheral vision, which is due to my squint. When I'm forced to focus with my left eye (it's my "weak eye") the vision is OK.
1	I have never been able to see well out of the affected eye.
1	I wear glasses so I think it is just normal vision problems. I don't think it has anything to do with the Rombergs.
1	Last year I went to see a eye doctor because my vision was getting bad. He found out that I had a detached retina and performed surgery to fix it. It has been good since then, but have noticed a slight blur in the past few weeks.

Continued on the next page.

Count	Page 28. III. 4. Comments (continued)
1	It blurs when reading from her eye movement problem.
1	Legally blind in affected eye.
1	Legally blind in affected eye. Ocular pressure extremely low (runs between 3 and 5). Eyelid does not close all the way.
1	Legally blind in left eye. Last exam vision was 20/1100.
1	Loss of vision and cataract on left eye (Romberg side), right eye perfect and needs no correction of sight.
1	Loss of vision over four days. Bascom Palmer Eye Institute diagnosed Parry Romberg Syndrome at age 23.
1	Minocular duplicity (double vision in one eye), plus loss of vision.
1	My vision is blurred and I don't have peripheral vision.
2	N/A
1	Not sure
1	Only temporary vision loss. It was corrected.
1	Poorer vision in right eye.
1	Slight loss of vision that is corrected with glasses.
1	Some decreased vision after cosmetic surgery.
1	Temporary loss of vision during stroke. Afflicted with "black blobs". Problem went away on it's own... a mystery to doctors.
1	The loss of vision is greater in the affected eye (left).
1	Vision gradually diminishing.
1	Vision in affected eye is dramatically different than the other, but is fine with eyeglasses. The optic nerve is atrophied, disc shaped. I have had field of vision tests for years and it seems that it has not changed since I was 30, am now 55.
1	Vision only altered if pupil dilated.
1	Wear glasses but can't recall at what age I started wearing them.
1	When left eye is closed, I have almost no vision in the right eye.

Number responding = 32 / percent responding = 22% of total respondents (143).

Page 29. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 2 of 2)

1. Please give the results if the affected person had surgery to correct a receding eye:

Count	Comments (20)
1	After I had filling material brought into my eyelids (natural fat and chemical fat) I could close my eye better and it became smaller (more as the other eye). But the positive effect receded after a few years.
1	All ok now. Dr. used bone chips that he got from my scalp to rebuild my eye socket as it had atrophied. This brought my eye up and forward to be more even with the other one. The Drs. also worked on eye lid so that eye isn't so dry anymore. It's not perfect, but it is much better than it was.
1	Botox injections
1	Fair
1	Fair- has a gold weight on eyelid- though needs a heavier weight placed.
1	First two fat transfer reabsorbed different surgeon - two fat injections some improvement I think if i had not had the first two, it would have had a better result.
1	Getting surgery soon to correct the eye.
1	I had skin grafts -- the tissue was taken from my left hip and placed behind my left eye.
1	I have not had this surgery. I understand it is very serious surgery because it effects eye vision and I would not be a suitable candidate.
1	I was told before the PRK that I was going to need total reconstruction of my eye orbit, temple and upper check to raise the left side orbit to be even with the right and bring my left eye forward. I declined this operation and saw four other doctors. One said the same thing as the first and the other three told me I did not need that and that I qualified for PRK surgery.
1	No - just fat injected under eye which gives the appearance of it not being so droopy. Still weeps alot.
9	No surgeries

Number responding = 20 / percent responding = 14% of total respondents (143).

Page 29. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 2 of 2)
2. Please describe any factors that caused dilation of the eye:

Count	Comments (13)
1	After every operation I had, my left eye will dilate and it will stay that way for a long time, then it will go down with some dilation left, but it never would go back to normal.
1	My daughter has a coloboma which is a uneven shaped pupil which does not get smaller in light
1	Nerve damage, right eye does not respond to light.
7	None
1	Not sure
1	Stabismis surgery caused the pupil to dilate fully and stay that way.
1	Unknown

Number responding = 13 / percent responding = 9% of total respondents (143).

Page 29. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 2 of 2)
3. How long did the dilation last?

Count	Comments (11)
1	2-4 weeks at a time
1	Continuing / unreturnably / irreversible
1	Dialation is continuous.
1	Dialation is still in effect.
1	For a long period of time, 2 or 3 months.
1	It has been ten years.
4	n/a
1	Permanent dilation of the left pupil

Number responding = 11 / percent responding = 8% of total respondents (143).

Page 29. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 2 of 2)

4. What measures did the affected person take to correct problems with dry eyes:

Count	Comments (31)
1	Best result with an eye spray to the eyelid. Avoid bright sunlight, keep blinds closed and wear sunglasses outside. Stay hydrated - drink plenty of water. Stay out of wind.
1	Dr. gave me samples, which I have not used.
7	Eye drops
1	Eye drops- see above for rebuilt eye socket and eyelid revision surgery.
1	Eye drops such as poly gel and Luxyal
1	Eye ointment every night
1	Had tear duct sealed which has not helped at all... use drops all the time.
1	I consciously blink more. I haven't discussed it with my doctor as yet, so don't have any drops at this stage.
1	I continually use drops for dry eyes.
2	I lubricate the eye with ointment and tear drops.
1	I put ointment in every night before sleep, and lubricating drops in a few times throughout the day.
1	I take supplements (GLAs, borage oil) which seem to help a lot, all the pain is gone, and I also put tear drops in my eye. At night I tape that eyelid shut.
1	I use an ointment called Lacrilube at night to keep my eye moist. If dryness or irritation occurs during the day, I use Refresh tear drops.
1	Lubricating eye drops and eye gel. I also wear a bandage contact lens on that eye.
4	N/A
1	Nightly use of PM refresh ointment.
1	Not sure
1	Over the counter eye drops at night before bed and in the morning.
1	Tear replacement during the day and stronger Rx at night.
1	The eye on the Rombergs side does not tear at all. I've been using Restasis which makes my eye glassy when I cry but it still doesn't tear.
1	They opened the tube next to my nose a bit, but it did not work. Other than that nothing was done.

Number responding = 31 / percent responding = 22% of total respondents (143).

Page 29. III. Associated Symptoms (page 2 of 4) cont.: Eye affected (page 2 of 2)

5. Comments concerning how the eye is affected:

Comments (23)
My eye gets infected often.... it hurts all the time.... the surgery caused my eyelashes to grow inside the eye area which irritates it.
Tight, does not rotate properly, double vision due to recessed eye.
Some drying of the eye at times due to the eyelid not completely closing during sleep.
There is almost continuous strain in the left eye now, causing pain and redness. I have to stop reading after a couple pages to rest the eye before starting to read again. Have not found out why this is yet.
Lower vision in rombergs eye (right), also extremely sensitive to light as only have partial eye lashes.
Weeping eye Blurred vision
The left eye protrudes, causing dryness. The right eye overcompensates for the dry left eye and waters all day long.
Did try steroid drops at one point, but not for long.
This eye was severely near-sighted (-8.5). This was corrected with lasik surgery about 10 years ago.
n/a
Droopy eyelid
Just asymetrical with the other eye, vision, etc. perfect. Hyperpigmentation around the left eye.
Eye does not close completely due to the lower lid being pulled down.
Tearing
I think the dry eye and episcleritis are due to the eye receding slightly and the eyelid not being able to close completely when sleeping.
Mitchell's eyelid dosen't close properly when he sleeps.
Avoid wind and getting salt water in eye - use drops.
Without having had the surgeries to correct the problems I had, I wouldn't be very happy- they were really bugging me, and I'm sure that my eye would be all good and scratched up by now!
Dry and gritty. Vision gets worse as it gets drier.
Not sure, but she has vision problems.
It is very annoying. I have to wear make-up because of my red skin, but when I am outside and get wind in my face, all the tearing washes away the make-up. In heated or cooled rooms I always get the opposite: dry eye. Sometimes I cannot do anything else but close my eyes.
I don't know if its associated with the Rombergs or not...............But i get these flare ups of itchy eyes, well the skin around my eyes.................they swell and can burn.
Right eye seems much smaller than left

Number responding = 23 / percent responding = 16% of total respondents (143).

Page 30. III. Associated Symptoms (page 2 of 4) cont.: Face Affected
1. Please select how the face is affected:

Affected	Responses (72)	% of Number responding
Facial pain	47	76%
Facial paralysis or weakness	25	40%

Number responding = 62 / percent responding = 43% of total respondents (143).
Note: This was a multiple choice question.

Note: If "Facial pain" was selected at the above question 30. III. 1. "Please select how the face is affected" then the following section (Page 31) would apply.

Page 31. III. Associated Symptoms (page 2 of 4) cont.: Face Affected
1. Describe the location of this Facial pain:

Responses (43)
above lip, jawline, head
all of the affected side
All on the affected side from the head to the jaw bone.
Anywhere along the line of the indentation. Also, in the left cheek (wondering recently if there is a problem with the left sinus).
cheek and upper lip
cheek area, eye, ear
Down side of face close to ear and temple, shooting pain up the side of my face.
During headaches, affected face part is sensitive.
extreme sharp pain in jaw jaw spasms
eye and under check bone
facial pain on Romberg side only
forehead twitching, weakness when chewing for long periods
forehead, around nose, and above lip
forehead, eye area
forehead, eye and nose area
Happened in my teens and 20s, especially tight jaw.
in cheek, jaw, it aches, throbs
In my cheek bone area down into my jaw, but it is not a constant pain for me.
It affects the whole area but not all at the same time.
It's like having headaches like something is growing... or sometimes like I've been hit with something in the face, it's not in fixed points, it's all over, the pain radiates.

Continued on the next page.

Page 31. III. 1. Responses (continued)
jaw
Its not really pain, but more odd sensations which are around the nose and under the eye and spread towards the ear on the left side....sometimes radiating down towards the mouth and chin. I don't know the answer to the next question, so I'm gonna leave it blank!!
jaw - TMJ cheek area neck pain on affected side 'pain' in teeth migraines sensitive scalp - mostly where the 'dent' is
jaw pain and discomfort
left temple, cheek
mandible, muscle spasms
mild pain on the cheek occasionally
My whole head is hurting if I lay on it or put it to my hand when I am writing or reading for example. And it hurts all over if it is cold.
not sure
numbness on side
Occasional facial pain occured during early years of condition - described as headache - very intense pain 'shooting' down left cheek area.
Occasional pain mainly in cheek area.
on the right jaw line
pressure like someone is pushing on it and hurts from the cold
right side
right side of the face - at first it was taken for TMJ pain
scalp on the area of lost hair forehead chin
Sometimes pain on forehead and jaw area of affected side.
Started as trigeminal neuralgia. Would go on and off. Ignored it for awhile, then was told I probably was grinding my teeth. Noticed jawline changing a bit on right side where pain had been. Have had pain in chin area where current changes are.
tingling and numbness
top of cheekbone to ear
Very short and sharp and on many different parts of the face and head - not debilitating like a migraine. Also get pain in the tongue.
whole right side of face

Number responding = 43 / percent responding = 30% of total respondents (143).

Page 31. III. Associated Symptoms (page 2 of 4) cont.: Face Affected
2. Is the pain triggered from the trigeminal nerve?

Triggered	Responses (25)	% of Number responding
Yes	18	72%
No	7	28%

Number responding = 25 / percent responding = 17% of total respondents (143).

Page 31. III. Associated Symptoms (page 2 of 4) cont.: Face Affected
3. Any medication or procedure that stopped this facial pain:

Count	Responses (24)
1	Acupuncture has completely corrected facial pain and muscle movement is back.
1	Analgesics periodically during that phase.
1	I can manage without medicine so far.
1	I do not know the answer to number two above. I am not aware of any medications that stop the facial pain. Usually it is a discomfort rather than a pain. My daughter uses over the counter Advil or Tylenol. Re: #5 below - it varies from very mild to moderate. That is my understanding as her mother.
1	I have adopted acupunture successfully.
1	I just use ibuprofen for mine it is a very mild pain.
1	I take Lortab which makes it tolerable.
9	no
1	Not at this stage.
1	Not severe enough to need any.
1	not sure
1	Nurofen
1	over the counter
1	Tamal Pax Paracetamol Voltarin
1	Tylenol
1	Vicodin can take the hard edge off the pain. Darvocet doesn't work as well as Vicodin. I think the pain runs its course, irrespective of pain meds. One of the difficult things about the pain is that there's no way to know when it will come on, how severe it will get, or how long it will last (anywhere from a few minutes to a week, or longer in rare incidences). There can be a reprieve from pain for a couple hours to a couple weeks. I never know how this will be. I have noticed in the past couple years that there does seem to be a link with ovulation (and sometimes menses) and 'dent' headaches. This is almost always the case. Although the onset of 'dent' headaches are not limited to ovulation. I have Poly- Cystic Ovarian Syndrome and didn't start having regular cycles until two years after my son was born. I'm somewhat new to the world of regular menstrual cycles and that's why it's taken me this long to figure out this pattern. Another correlation I've made the past couple years is that of weather changes (atmospheric pressure) could be a factor in onset of 'dent' headache pain.

Number responding = 24 / percent responding = 17% of total respondents (143).

Page 31. III. Associated Symptoms (page 2 of 4) cont.: Face Affected
4. Please comment on effectiveness of this medication or procedure.

Medication or Procedure	Responses (9)
Acupuncture has completely...	Acupuncture and acupressure were/are extremely effective for muscle movement and pain. We can consider it back to "normal."
Vicodin	Effective in taking the hard edge off the pain, but does not eliminate it.
Nurofen	gives some relief
Analgesics	Good effect - this type of pain occured at irregular intervals and has since abated.
No	I don't have any comments.
Ibuprofen	It works for me.
Tamal	Medication works for a while and then pain returns.
Tylenol	N/A
None	none

Number responding = 9 / percent responding = 6% of total respondents (143).

Page 31. III. Associated Symptoms (page 2 of 4) cont.: Face Affected
5. Rate the severity of this facial pain:

Rating	Responses (43)	% of Number responding
1 (very mild)	3	7%
2 (mild)	17	40%
3 (moderate)	17	40%
4 (severe)	5	12%
5 (very severe)	1	2%

Number responding = 43 / percent responding = 30% of total respondents (143).

Page 31. III. Associated Symptoms (page 2 of 4) cont.: Face Affected
6. Comments for Facial Pain:

Responses (17)
can be very aggravating
Cold weather is very painful. Day to day there is a sensation of 'tightening'-slight pain.
Comes and goes. Somtimes worse than others.
Facial pain is anywhere from mild to very severe. It is a major factor in the quality of my life.
Have had surgery so these can also be from that. I don't recall any pain prior to surgery.
I don't know what nerve tiggers it. It comes in bouts. I may not get it for months, then I get it for 3 days in a row! Sometimes it seems to be connected to stress, but sometimes it just comes on when I'm not feeling stressed at all! It also sometimes comes on when I eat nuts or tough food (which I try to avoid now).
I try to take little notice of it and give it a rub now and then. I can't talk on the phone for long using the right ear as my face just burns!
It comes and goes, makes it worse by the biting of the jaw.
It would come and go.
moderate but frequent
not sure
Pain generally constant. Seems worsened by fatigue.
Pain may be due to microvascular free flap procedure.
Pain only occasional, when I'm overly tired.
See above. These questions are difficult to answer. Sometimes you don't have a choice that fits best.
The pain occurs in unregular periods. In this pain-period the severity is moderate to severe (touching the affected area makes the pain worse).
Usually I have it when I'm in my best moment, it's starts suddenly and that way stops too.

Number responding = 17 / percent responding = 12% of total respondents (143).

Note: If "Facial paralysis or weakness" was selected at the above question 30. III. 1. "Please select how the face is affected" then the following section (Page 32) would apply.

Page 32. III. Associated Symptoms (page 2 of 4) cont.: Facial paralysis or weakness

1. Do you feel this facial paralysis or weakness is related to Rombergs?

Related	Responses (24)	% of Number responding
Yes	22	92%
No	2	8%

Number responding = 24 / percent responding = 17% of total respondents (143).

Page 32. III. Associated Symptoms (page 2 of 4) cont.: Facial paralysis or weakness

2. Comments for facial paralysis or weakness:

Responses (11)
bite is not as strong in affected side
due to surgery
Gradual onset age 19-22 years, the only symptom of PRS I had at that age and the reason I was first diagnosed.
I only get it on the left side of my face.
I think there is some damage caused by my surgery and some caused directly from the PRS still being active.
mouth droops down on left side, smile no longer symmetrical
My facial paralysis could be a mild case of cerebral palsy due to premature birth - have had differing opinions on this.
My right side is weaker then my left side. If I talk for extended periods of time I find it difficult to move my jaw to form words. When I puff my cheeks out I am unable to puff the right side out as much as there doesn't feel like there is any muscle strength there.
She has problems with her jaw and severe deterioration of affected areas.
The eye muscle is paralyzed and does not allow the eye to look to the right, therefore I have to move my head to compensate.
The weakness was around the mouth where muscle had atrophied.

Number responding = 11 / percent responding = 8% of total respondents (143).

Page 33. III. Associated Symptoms (page 2 of 4) cont.: Fasciculation (muscle twitching)

1. Can it be linked to the start of progression:

Linked	Responses (35)	% of Number responding
Yes	19	54%
No	16	46%

Number responding = 35 / percent responding = 24% of total respondents (143).

Page 33. III. Associated Symptoms (page 2 of 4) cont.: Fasciculation (muscle twitching)

2. How long did it last:

Count	Responses (27)
1	?
1	10 years
1	15 years
1	19 - 27
1	6-12 months, best guess
1	a few seconds
1	about 13 years
1	almost 4-5 years
1	comes and goes, maybe last a day or a few days
1	eyelid twitching comes and goes
1	I still have it, amount of time it lasts varies in seconds.
1	It used to last just minutes.
1	mild and brief
1	not long in my case
2	not sure - it comes and goes
1	Not sure- didn't really keep track- it has just been something that happened.
1	off and on for 3 years
1	on and off
1	rarely occurs
1	since it was diagnosed at age 15
4	still going on
1	still occurs occasionally
1	unknown

Number responding = 27 / percent responding = 19% of total respondents (143).

Page 33. III. Associated Symptoms (page 2 of 4) cont.:
Fasciculation (muscle twitching)
3. Can it be linked to a stage of activity:

Count	Responses (21)
1	?
1	For me - possibly linked to beginning stages.
1	I don't know
1	I think with the time when the atrophy started.
5	no
1	none that I can think of
4	not sure
1	not that I know
1	seems to happen more when I am fatigued
1	unknown
3	yes
1	yes after the blunt trauma

Number responding = 21 / percent responding = 15% of total respondents (143).

Page 33. III. Associated Symptoms (page 2 of 4) cont.:
Fasciculation (muscle twitching)
4. Comments for Fasciculation (muscle twitching):

Responses (22)
Can't recall twitching prior to surgery in '89.
Doesn't happen all that often, maybe a few times every few weeks. Even so, it isn't always in the affected areas, or even the affected side.
eyelid twitching, came late as it reached my eye area
happens in the spur of the moment
Happens rarely, but I do feel like the deterioration is taking place while it is happening.
I also had jaw lock due to spasm of muscle in jaw. Muscle would harden like a nut - for several minutes. Sometimes back to back a number of times. Eventually, muscle would tire out and it would be painful to open mouth at all.
I did not have this before the surgery.
I do get some, but I do not know if it is associated with progression.
I get twitching around my left eye sometimes. I haven't thought about the timing previously.
I remember that it started happening to me just before I had my free-flap surgery, and I freaked out and called Boston to let the Dr. know.... he wasn't concerned, so I figured it wasn't a big deal. I thought that my nerves were dying or something!!! Every now and then it happens now and I'm like, uh oh.... here we go again, but I don't worry about it.
I've had slight muscle twitching in the affected side of my face for years. For several months now I've started to have muscle twitching throughout my body. Will be seeing a doctor about it this next month (may not be related to Rombergs).
Meds did not seem to help, only the induced coma would stop them, Ativan was helpful at times.
Mine has been with me all my life since it was diagnosed at age 15.
Neck and left shoulder feel weak and I find it hard to control the twitching, especially when concentrating or driving.
no set pattern noticed
none
not severe
Still get twitching in jaw and cheek. Haven't had the pain for about 2 yrs. Still get migraines, but not as often as before.
The fasciculation is the locking of my jaws.
The muscles twitch in the face and on my arms and legs.
very annoying
very annoying, cold weather really bothers the left side of my face

Number responding = 22 / percent responding = 15% of total respondents (143).

Page 34. III. Associated Symptoms (page 2 of 4) cont.: Forehead affected

1. Please select how the Forehead is affected:

Affected	Responses (169)	% of Number responding
Bone loss	25	36%
Discoloration	28	41%
En coup de sabre (indentation)	58	84%
Protruding bone	5	7%
Thin skin	45	65%
Other (Specify)	8	12%

Number responding = 69 / percent responding = 48% of total respondents (143).
Note: This was a multiple choice question.

Page 34. III. Associated Symptoms (page 2 of 4) cont.: Forehead affected

1. Please select how the Forehead is affected:

Other (Specify)

% of Total Responses	Responses (8)	Other - How the Forehead is Affected
0.6%	1	atrophy
0.6%	1	hair loss along the entire line of it.
0.6%	1	shiny skin
0.6%	1	slight atrophy right temple
0.6%	1	slight tissue loss
1.2%	2	tissue loss
0.6%	1	VERY TENDER TO TOUCH AROUND THE EYE

Page 34. III. Associated Symptoms (page 2 of 4) cont.: Forehead affected

2. Comments for Forehead affected:

Responses (19)
I have it mildly on both sides of my forehead.
Not as bad as the cheek.
I feel that the loss of subcutaneous tissue has made my bone appear to be protruding.
bone graft age 17
The protruding bone is in the area next to the indentation, so it just looks like its protruding. The skin is shiny but not discolored.
mild en coup de sabre.... thin skin in temple area
The skin on my forhead is extremely thin, the color is darker and the indent is severe, but all confined to the right side.
Two lines running down to eyebrow, slightly indented slightly pigmented. The rest of my left forehead has very thin skin so the blood vessels are visible through it. The skin has no visible pores or acne (the rest of my face does).
Sometimes the line does look white, other times bluish - so discoloration might also apply - her CT scan it appeared that you could slightly see the indention.
Left side of forehead is normal, right side is flat.
distinct lines or stripes
Left side of forehead has small indentation from Romberg's. I have had fat injections done and it helped out alot.
As I am rather overweight, it is very noticable that it is now just skin on bone.
small indent in tissue loss
I also have a dent above my eye brow and a receding hair line on the left side.
Not sure about bone loss... I think so?
only slightly and very recently
looks like dent
have had several surgeries to help correct this

Number responding = 19 / percent responding = 13% of total respondents (143).

Page 35. III. Associated Symptoms (page 2 of 4) cont.: Hair affected

1. Please select how is the Hair affected:

Affected	Responses (118)	% of Number responding
Alopecia (hair loss)	39	71%
Excessive hair	0	0%
Eyebrow (missing all or part of)	24	44%
Eyelashes (missing all or part of)	22	40%
Loss of body hair	2	4%
Thinning	23	42%
Other (Specify)	8	15%

Number responding = 55 / percent responding = 38% of total respondents (143).
Note: This was a multiple choice question.

Page 35. III. Associated Symptoms (page 2 of 4) cont.: Hair affected

1. Please select how is the Hair affected:

Other (Specify)

% of Total Responses	Responses (8)	Other - How is the Hair Affected
0.8%	1	bald spot
2.5%	3	gray hair
0.8%	1	receding hairline
0.8%	1	streaks of gray hair as baby
0.8%	1	vitiligo
0.8%	1	whiter on right side

Page 35. III. Associated Symptoms (page 2 of 4) cont.: Hair affected

2. Comments for Hair affected:

Responses (25)
Not sure if it is related to hypothyroidism.
Hairless patch extending midway back on scalp, alongside of en coup de sabre.
I had parts of the head without hair but I had cut it all and after that made a treatment with carbonic snow and grew almost 70% back.
Hair loss is only along the line of the indentation.
My hair is thinner on affected side and a small area is completely white. No big problem.
It follows right up the line of the dent on my forehead.
She has had gray hair only on the left side. It has to be colored to hide it.
The hair in the temple area has receded and thinned. Some is just gone.
Hard to tell if high forehead is due to Romberg's or just a family trait...my father had a high forehead/temple line, but no Romberg's.
I had a small amount of hair loss at age 15 but then it stopped. About 2 years ago it started up again and now I have lost the majority of my hair on the right side, about 3 inches wide from my forehead to the back of my skull. It follows the coupe de sabre.
Afterwards I can say that since (or just before) it started, I had one spot on my head where the hair was completely white. Everybody laughed about it and said that I started young to become gray. However, I did not got more gray hair beside that one spot. Now I'm 45 I'm getting a bit more yes. :P
She has the main indention, then a smaller one about 1" away from the main one - both have hair loss (the main indention has the most hair loss).
gray hair in scalp, eyebrows and eyelashes
Hair grows unevenly.
Alopecia in the affected areas… they were totally bald but there is now some regrowth but the hair is very thin in thickness.
There is a bald spot on the right side of his head that was there when he was born and it has unusual skin in that spot also almost like a burn.
Right side of face ---- hair line is receding.
In the beginning there was a small area without hair - ca 1 cm2 now (within 20 years) this area has grown to ca 5 cm2.
Not having any hair on the en coup de sabre really was hard while being a teenager, and young adult... especially when swimming or when the wind would blow, etc!! Now, that the Dr. has fixed it, no worries, but the flat spot on the back of my head is bald, and always has been- the other day, my daughter commented on it while waiting for the bus... not that I worry about it now, but it is a hard part of this disease - especially for the young people!
hair loss at the left temple part of lower eyelashes missing medially
The right side is thinning more than the left particularly around the temple area.
hair is gray
mild - small spot missing hair
hair loss - en coup de sabre?
We thought it was caused by eczema or allergies when she was younger. Hair loss was actually before 15, like 10, but the surgery was at 15.

Number responding = 25 / percent responding = 17% of total respondents (143).

Page 36. III. Associated Symptoms (page 2 of 4) cont.: Headaches
1. Please select the type/types of Headaches:

Affected	Responses (145)	% of Number responding
Fatigue	33	54%
Migraines	52	85%
Muscle weakness	14	23%
Nausea	17	28%
Slurred speech	9	15%
Vomiting	12	20%
Other (Specify)	8	13%

Number responding = 61 / percent responding = 43% of total respondents (143).
Note: This was a multiple choice question.

Page 36. III. Associated Symptoms (page 2 of 4) cont.: Headaches
1. Please select the type/types of Headaches:
Other (Specify)

% of Total Responses	Responses (8)	Other - Type/Types of Headaches
0.7%	1	dizziness/vertigo
1.4%	2	light sensitivity
0.7%	1	normal headache
0.7%	1	occasional vomiting, sensitivity to light and sound
0.7%	1	sensitivity to light and noise
0.7%	1	sinus
0.7%	1	tension

Page 36. III. Associated Symptoms (page 2 of 4) cont.: Headaches

2. Comments for Headaches:

Comments (28)
I would not say they are migraines- sometimes I have nausea associated with the headache sometimes I am sensitive to light and sound. They are normally located in the front part of my head and eyes.
Seem to always be there, though can be more or less severe.
I got migraines that lasted for months.
When I first got hit in the jaw when I was 8 years old I remember that night being in severe pain. I had such a migraine that I felt debilitated and very sick. The next day I was better but that's when the blue spot appeared on my jaw. Every 4 years I would get this same migraine pain and afterward I would get electical impulse twitches, then it would fade out and go away. But after this migraine I would see facial changes for a period of 4 years then it would burn out.
Not positive headache caused by Rombergs, due to allergies, it could be one or both ailments.
Sometimes sharp pains, sometimes dull ache.
The sinus headaches may or may not be related to Rombergs, but I believe that my sinus cavities are affected by the atrophy on the left side of my face.
They could last for a few seconds or minutes and sometimes hours....... it's not constant and not in one place..... it's a feeling like someone would hit you with a sharp and thin object like a very long needle.
I have headaches daily now. I think that this has to be the worst part of it because I am so uncomfortable.
I don't know that I would classify it as "migraines." Although it is confined to the affected side of my head (left) the pain is always within the area of indentation, not the whole left side of the head. It feels like a hard pressure and/or pulling apart in the indentation. There's also sometimes a sharp/intense pain in my left cheek. I suppose this could be along the line of the indentation, and maybe is part of that, or it is the sinus cavity that's affected somehow.
It seems she gets more frequent headaches when it's more active.
Excruciating pain when in childhood and as adolescent.
My headaches get so bad that there are times I need to be taken to the ER for treatment. Other times Tylenol helps.
She has only complained of one headache that could have possibly been a migraine, the rest were what I call her regular headaches, managed with medicine and once it has taken effect she can get back on track.
Has headaches at least twice a week. Usually controlled by 1 or 2 doses of Tylenol. Follow up with Neurology.
Chronic headaches and migraines. I see a neurologist and take migraine prevention medication.
I have slurred speech especially when tired.
I used to get migraines and headaches a lot when I first got PRS. Now I only get 1 or 2 migraines a year and probably a headache every one or two months. Most of my migraines were the painless ones where I had problems with my sight, e.g. auras, slurred speech where I can't think of the words then can't get them out and I lose control of my right arm. I also lose control of my right arm when I am really upset about something or very angry.
light sensitive, noise sensitive
I assume they are related to Rombergs, but it is hard to tell for sure.
Unregularly periods of headaches, only on the right side of the head with great pain around and behind the eye.
NO fun!!! I used to get headaches nearly every day- I guess they could be attributed to being tired- I worked on the night shift back then, and two young children.. or stress...young children again..... not sure, but in 2004, I had the absolute worst headache ever on the right side - felt like a knife was being stabbed through and I asked my husband to take me to the emergency room which he did- they didn't do anything, but soon after my Neurologist did an MRI, and found lesions... go figure!
have eased up significantly
After headache started, takes Diclofenac to reduce pain. Fall asleep after a few hours and sleep for several hours. That day still very tired because headache costed lot of energy.

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Page 36. III. 2. Comments (continued)
Its not just on my affected side, but in my whole head.
I always have a headache... it just how bad it is going to be today. Tried lots of meds and Motrin 800 is what works for me.
tension headches
We have to watch what she eats, no nutrisweets, chocolate, diet pop, no caffeine, no nitrates, no MSG, watch the sun, watch how much she reads.

Number responding = 28 / percent responding = 20% of total respondents (143).

Page 37. III. Associated Symptoms of Rombergs (page 3 of 4)
1. Please select all that apply (page 3):

Symptoms	Responses (319)	% of Number responding
Heart affected	1	0.9%
Jaw affected	59	56%
Joint affected	22	21%
Morphea	24	23%
Mouth affected	59	56%
Muscle loss	40	38%
Nose affected	55	52%
Numbness	31	30%
Problems with balance	28	27%

Number responding = 105 / percent responding = 73% of total respondents (143).
Note: This was a multiple choice question.

Page 38. III. Associated Symptoms (page 3 of 4) cont.: Heart affected
1. Please select how the Heart is affected:

Affected	Responses (1)	% of Number responding
Ailments	0	0%
Mitral valve prolapse with regurgitation	0	0%
Murmur	1	100%

Number responding = 1 / percent responding = 0.7% of total respondents (143).

Page 38. III. Associated Symptoms (page 3 of 4) cont.: Heart affected
2. Comments for Heart affected:

Count	Responses (0)

Number responding = 0 / percent responding = 0% of total respondents (143).

Page 39. III. Associated Symptoms (page 3 of 4) cont.: Jaw affected

1. Please select how the Jaw is affected:

Affected	Responses (293)	% of Number responding
Atrophy of the jaw bone	35	61%
Bite that feels uncomfortable	23	40%
Biting of the tongue or cheek	22	39%
Inability to open comfortably	24	42%
Grinding teeth	18	32%
Jaw clicks	24	42%
Jaw locking	19	33%
Jaw spasms	22	39%
Limited opening	26	46%
Misalignment between upper and lower jaw	30	53%
Overbite	10	18%
Pain in the jaw joint	20	35%
TMJ disorder (Temporomandibular Joint in Jaw)	13	23%
Other (Specify)	7	12%

Number responding = 57 / percent responding = 40% of total respondents (143).
Note: This was a multiple choice question.

Page 39. III. Associated Symptoms (page 3 of 4) cont.: Jaw affected
1. Please select how the Jaw is affected:
Other (Specify)

% of Total Responses	Responses (7)	Other - Jaw Affected
0.3%	1	atrophy of tissue/jowl
0.3%	1	loss of teeth
0.3%	1	major dental problems with teeth
0.3%	1	narrow Jaw, teeth crowding
0.3%	1	only visual
0.3%	1	really small mouth
0.3%	1	un-sure of all her problems

Page 39. III. Associated Symptoms (page 3 of 4) cont.: Jaw affected
2. Please describe the steps that were taken to resolve any of these problems:

Count	Responses (32)
1	mouth guard at night
5	none
1	I had braces put on when I was 14 years old, I had them for 5 years. I still have all the problems stated above. I just recently had my wisdom teeth removed but only for impaction. My wisdom teeth did not try to come in until I was 24 years old and they where very very painful.
1	HAD RETAINER - WORN OVERNIGHT FOR A PERIOD OF ALMOST A YEAR WHEN AGED 11 - 12.
1	I have a botox needle every four months to stop the spasms in the jaw.
1	Regular trips to hospital to have jaw cleaned? (This did not happen until after an operation to have skin etc. taken from my scalp to fill out my chin and nose) which included being put out to have jaw opened wide etc. I stopped this several years ago when I was told that PRS had definitely stopped and now survive with the use of 2 very good mouth supports which I use when my jaw locks or goes into spasms.
1	Went to the dentist, gave me a mouth piece to use during sleep.
1	The doctors did not help me with anything. I did not receive treatment or help for it
1	reconstructive surgery to jaw and teeth
1	pain medication
1	She has had braces and reconstructive jaw surgery, Leforte osteotomy, leveled upper jaw, sagittal split osteotomy, very small mandible, moved bone from one side of chin to other.
1	With braces and the free flap, many of these symptoms alleviated.
1	I have not seen a dentist in a few years because none that I have seen know anything about PRS and they force my jaw to open. They have all said it's in my head and if you really wanted to open your mouth you could.
1	Removal of affected gum meant loss of two front teeth, bow fitted with partial plate. surgery fixed some things now in later life teeth are a problem. the rest just live with

Continued on the next page.

Count	Page 39. III. 2. Responses (continued)
1	physiotherapy
1	Mouth guards for TMJ and appliances for opening upper and lower jaws.
1	physical therapy
1	I no longer have jaw clicks, locks or spasms. However, I did - they seem to be stages of my progression. I have 18 crowns - to correct the misalignment between my upper and lower jaw. I had a completely open bite on my affected side prior to my crowns.
1	Tried muscle relaxants when jaw spasms severe but not successfully.
1	Occipital plate - worn every night to prevent clenching, lessen the pressure on the TMJ. Physiotherapy - once and never needed it again, jaw also stopped clicking.
1	Teeth on top were arched. Bones were cut and moved and filled in the space with bone graft and a bridge.
1	Right now nothing. She needs braces, teeth and bone implants. Missing a lot of permanent teeth on left side. Teeth are very tiny.
1	No surgeries done on the jaw. The dentist did put something on my teeth so that they would touch each other (upper and lower jaw) but it did not work.
1	face lift
3	braces
1	None yet. She needs braces, bone and teeth implants but not until she is 16 - 18 yrs. old. Missing most of left side top and bottom permanent teeth. She is compared to a 70 yr. old's mouth.

Number responding = 32 / percent responding = 22% of total respondents (143).

Page 39. III. Associated Symptoms (page 3 of 4) cont.: Jaw affected
3. Please describe if any of these steps were successful:

Steps that were taken	Responses (17)
I had braces put on when I was 14 years old,…….	They helped straighten my teeth and my self esteem. I felt better afterwards because my teeth looked straight instead of having teeth that did not fit my mouth. I also had to have some teeth removed so my other teeth would have room to come in.
HAD RETAINER - WORN OVERNIGHT FOR…..	YES - AS SUCCESSFUL AS POSSIBLE
regular trips to hospital to have……	not fully
Went to the dentist gave me a……	A little but the jaw pain is being affected during the day, which you cannot go out in public wearing the piece.
reconstructive surgery to jaw and teeth	yes
Pain medication	no
She has had braces and reconstructive jaw…..	All was very successful.
Physiotherapy	Did not work.
mouth guards for TMJ and appliances for….	Yes, in both cases.
Physical therapy	It did help with enabling me to open my mouth wider.
I no longer have jaw clicks, locks or spasms. However….	So far the crowns have been successful.
tried muscle relaxants when jaw spasms……	Bone grafting and dental implants were successful
Occipital plate - worn every night to prevent….	Physiotherapy - once and never needed it again, jaw also stopped clicking.
Teeth on top were arched. Bones were cut…..	Best surgery out of all of them.
Braces on her teeth twice.	not sure
face lift	face was made symmetrical
Braces	very successful

Number responding = 17 / percent responding = 12% of total respondents (143).

Page 39. III. Associated Symptoms (page 3 of 4) cont.: Jaw affected

4. Comments for Jaw affected:

Responses (16)
comes and goes
I believe my jaw is progressively getting worse, potentially from PRS and potentially from normal wear and tear on an atrophied jaw. To date I have not sought treatment on it, but due to recent episodes of pain and discomfort I will have to in the future.
It's very disturbing and it limits my actions.
Since jaw affectation is mild, I've never sought remedy...I just live with it.
The atrophy in the jaw bone is mild.
I have a perforated disc in the TMJ and have to be careful when chewing food.
This is embarrasing and hurts alot.
Can not open my mouth comfortably.
Surgeon is going to try to assist my mouth in opening further.
Get unexplained pains regularly.
Mild discomfort unless cold.
Serious deterioration of jaw and tissues of area, also color above lip and area of jaw is affected.
I cannot chew on the affected side. I always chew my food on my 'good' side. Unfortunately this does not help the muscles in my affected side.
Right side of jaw will click.
Tooth root loss is significant on right side. No teeth lost yet but dentist thinks it is inevitable.
Missing or thinning of jaw bone.

Number responding = 16 / percent responding = 11% of total respondents (143).

Page 40. III. Associated Symptoms (page 3 of 4) cont.: Joint affected
1. Which joint(s) is/are affected:

Count	Responses (18)
1	all - she developed arthritis
1	elbow and knee on affected side
1	hand joint pain, could be arthritis
1	hands, knees, hips
1	jaw
1	jaw / mandibular joint
1	jaw, cheekbone
1	left Jaw bone
1	left knee and elbow
1	left knee, hip, elbow, wrist, shoulder
1	left side joints, knee, hip, elbow
1	legs and arms
1	major joints… knees… elbows...
1	not sure
1	none
1	right ankle
2	TMJ (Temporomandibular Joint in Jaw)

Number responding = 18 / percent responding = 12% of total respondents (143).

Page 40. III. Associated Symptoms (page 3 of 4) cont.: Joint affected
2. Please rate the joint impairment:

Rating	Responses (20)	% of Number responding
1 (very mild)	3	15%
2 (mild)	2	10%
3 (moderate)	8	40%
4 (severe)	6	30%
5 (very severe)	1	5%

Number responding = 20 / percent responding = 14% of total respondents (143).

Page 40. III. Associated Symptoms (page 3 of 4) cont.: Joint affected
3. Comments for Joint affected:

Responses (10)
I have to exercise or else the pain is unbearable.
Only knee is severe. All others are mild so far.
I believe that PRS has weakend the muscles on the left side of my body, causing me to injure both my elbow and knee on the affected side of my body.
Symptoms alleviated with the surgery and braces... one cannot say if joint pain in other areas of the body are due to Rombergs unless a medical professional says so.
Lower jaw is uneven positioned.
Mostly noticeable during weather changes.
They click, grind, crunch when used/rotated. No pain.
She has arthritis along with loose joints. Weather plays a big part in how she feels. There are times she can barely walk.
not sure
Weather is permitted by how she feels. There are days she can not get out of bed. Then there are days she is just like a typical 8 year old.

Number responding = 10 / percent responding = 7% of total respondents (143).

Page 41. III. Associated Symptoms (page 3 of 4) cont.: Morphea

1. Please select the location of Morphea on the head:

Affected	Responses (59)	% of Number responding
Cheek	10	50%
Chin	10	50%
Ear	2	10%
Eye lid	4	20%
Forehead	9	45%
Lower lip	4	20%
Upper lip	4	20%
Nose	4	20%
Scalp	7	35%
Temple	5	25%

Number responding = 20 / percent responding = 14% of total respondents (143).
Note: This was a multiple choice question.

Page 41. III. Associated Symptoms (page 3 of 4) cont.: Morphea

2. Comments for Morphea on the head:

Responses (3)
I did not know I had on my head until the doctors found it. There was no pain only hair loss and there was a small atrophy but without a hard tissue.
Maybe I am confused but it would be the same as the En coup de sabre ??
If by morphea, you mean pigmentation changes, I've got them on my face. They started on the forehead and then on both sides of cheeks and jaw. Dermatologist said hard to distinguish between condition and sun damage, but since most of these have appeared since I've been wearing high SPF, don't know what to think.

Number responding = 3 / percent responding = 2% of total respondents (143).

Page 41. III. Associated Symptoms (page 3 of 4) cont.: Morphea

3. Other locations of the body with Morphea:

Other locations	Responses (26)	% of Number responding
Arm	4	40%
Back	7	70%
Chest	2	20%
Leg	4	40%
Neck	2	20%
Side	3	30%
Stomach	2	20%
Other (Specify) **Belly, Foot**	2	20%

Number responding = 10 / percent responding = 7% of total respondents (143).
Note: This was a multiple choice question.

Page 41. III. Associated Symptoms (page 3 of 4) cont.: Morphea

4. Comments for Morphea on other locations of the body:

Responses (1)
It's located only on my face and head.

Number responding = 1 / percent responding = 0.7% of total respondents (143).

Page 41. III. Associated Symptoms (page 3 of 4) cont.: Morphea

5. General comments about Morphea:

Responses (2)
My morphea is almost level with a slight indent with numbness and color loss. Dryness and no hair growth.
The same I found in the book.......... hard tissue..... colored skin.... pain..... a permanent ache sensation .... the pores of the skin are visible and bigger.

Number responding = 2 / percent responding = 1% of total respondents (143).

Page 42. III. Associated Symptoms (page 3 of 4) cont.: Mouth affected
1. Please select how the Mouth is affected:

Affected	Responses (134)	% of Number responding
Asymmetrical	42	71%
Palette	16	27%
Thinning lip	43	73%
Tongue	25	42%
Other (Specify)	8	14%

Number responding = 59 / percent responding = 41% of total respondents (143).
Note: This was a multiple choice question.

Page 42. III. Associated Symptoms (page 3 of 4) cont.: Mouth affected
1. Please select how the Mouth is affected:
Other (Specify)

% of Total Responses	Responses (8)	Other - Mouth Affected
0.8%	1	bone loss upper palette
0.8%	1	gums
0.8%	1	missing teeth in direct line
0.8%	1	not sure
0.8%	1	receding gums
0.8%	1	small mouth
0.8%	1	sunken cheeks - causes me to bite the inside of my cheeks, grind teeth
0.8%	1	teeth problems

Page 42. III. Associated Symptoms (page 3 of 4) cont.: Mouth affected
2. Comments for Mouth Affected:

Responses (19)
Mouth did not move on the PRS side when talking. Lower lip drawing up.
Tongue affected in that left hand side is smaller - atrophy noticable - lower lip is thinner on affected side.
Went to another consultant for the hole in the upper pallet. Said they could do nothing. I have to watch no food gets stuck into it.
Teeth very misaligned.
Right side, upper lip, thinner.
No problems eating, swallowing, or talking.
Root atrophy of upper teeth and bone loss on that side as well. I was told I would need a bone graft.
I 'grin' out of my right side.
Asthetically can't put lipstick on nicely, don't smile in photos, and in later life it is going to cause problems. Oh yeah kids ask if you been in an accident, adults assume you have a hare lip.
see last answers
I can tell when my PRS has been active as I bite the right side of my mouth while it is active until I get used to the changes and it stops. Have had some issues with choking on my own saliva during these periods too as swallowing is affected. Drinking out of some shaped glasses is also difficult as my lips don't go around the glass properly.
Teeth cantered
sharp pains in tongue quite often grind teeth at night involuntarily bite tongue and cheek - like a spasm
Atrophy on the right side of my tongue.
Everything seems thinner on the left.
Palette is higher in the left side. Top lip thinning on left side.
Bone loss resulted in loss of top teeth. Need a bone transplant.
She will give details when she completes survey, I am not sure.
Lip vanishing on left side............ My cheeks are sunken in and I bite my tongue alot.................... not sure whether its a tongue problem or cheek problem.

Number responding = 19 / percent responding = 13% of total respondents (143).

Page 43. III. Associated Symptoms (page 3 of 4) cont.: Muscle loss
1. Select the location of muscle loss:

Location	Responses (82)	% of Number responding
Abdominal	1	3%
Arm	12	32%
Back	5	13%
Chest	2	5%
Eye	11	29%
Jaw	23	60%
Leg	7	18%
Neck	13	34%
Other (Specify)	8	21%

Number responding = 38 / percent responding = 27% of total respondents (143).
Note: This was a multiple choice question.

Page 43. III. Associated Symptoms (page 3 of 4) cont.: Muscle loss
1. Select the location of muscle loss:
Other (Specify)

% of Total Responses	Responses (8)	Other - Muscle Loss
3.7%	3	cheek
1.2%	1	face
1.2%	1	face - right side
1.2%	1	jaw and cheek
1.2%	1	mouth
1.2%	1	not sure

Page 43. III. Associated Symptoms (page 3 of 4) cont.: Muscle loss

2. Comments for Muscle Loss:

Responses (9)
Can't be positive that the muscle loss in the arm and leg is due to PRS, but I don't have a similar pattern of muscle loss on the non-affected side of my body.
Now when the tissue it's not hardened and the atrophy "cleaned" all from my face and under my chin, sometimes I have the feeling that the parts that are missing are there like they contract or like pulsation, but in reality it doesn't happen. It's a very strange feeling.
No masseter muscle present on the left cheek.
Most of the loss of muscle tone in my body is due to not exercising regularly.
Due to lack of innervation on right side of face due to 7th cranial nerve palsy, left leg and left arm due to paralysis from brain. No muscle loss directly due to PRS.
Some atrophy on my right side of my neck internal and external. Loss of tissue and some weakness around my right elbow.
It's hard to tell the difference between muscle and fat sometimes! I suspect my abdomen also has less muscle.
If lifting weights, my muscle "power" is far less on the affected side.
I know of facial problems but not sure of the rest of body.

Number responding = 9 / percent responding = 6% of total respondents (143).

Page 44. III. Associated Symptoms (page 3 of 4) cont.: Nose affected
1. Please select how the Nose is affected:

Location	Responses (139)	% of Number responding
Asymmetrical	42	76%
Deviated septum	13	24%
Loss of tissue	31	56%
Pulled to one side	31	56%
Sinus cavity is thinning	17	31%
Other (Specify)	5	9%

Number responding = 55 / percent responding = 38% of total respondents (143).
Note: This was a multiple choice question.

Page 44. III. Associated Symptoms (page 3 of 4) cont.: Nose affected
1. Please select how the Nose is affected:
Other (Specify)

% of Total Responses	Responses (5)	Other - Nose Affected
0.7%	1	breathing
0.7%	1	just the very top between the eyes
0.7%	1	not sure
0.7%	1	very large sinus cavity on left side
0.7%	1	visible line

Page 44. III. Associated Symptoms (page 3 of 4) cont.: Nose affected

2. Comments for Nose affected:

Responses (15)
"Sinus cavity is thinning" is just a guess because of the pain in that area of the cheek only on the affected side—this has not been confirmed by a doctor.
Right side of nostril, thinner.
Cysts in sinus cavity.
The nose is thinner on affected side.
I had sinus surgery on the affected side, it has helped me breathe better but nothing cosmetic.
It gets cold on the tip. Plastic surgery, where a piece of my cheek was put on the tip to make it look more normal was a waste of time really cause it sunk in and after 15 years looking back it probably was better as it was, than it is now.
Just the very top of her nose between her eyes is slightly affected.
Has been experiencing nose bleeds and dry nose. Following up with ENT.
Visual line (coloration of skin change), looks like one side of nose is shrinking.
Loss of tissue on right side. The end hasn't been affected so the mid section which has lost the most tissues isn't as noticable.
Nose slightly fatter on one side.
No internal examination to date.
It feels like my sinus cavity is thinning, however, I have not been diagnosed with this.
Doesn't feel as if I can breath properly when I lay on affected side to sleep - have to breath through mouth. Can breath easily through nose when laying on good side.
She has lot of problems with right side of nose, inside and outside.

Number responding = 15 / percent responding = 10% of total respondents (143).

Page 45. III. Associated Symptoms (page 3 of 4) cont.: Numbness
1. Please select the location of Numbness on the Head:

Affected	Responses (95)	% of Number responding
Cheek	21	78%
Chin	11	41%
Ear	4	15%
Eye lid	5	19%
Forehead	11	41%
Lower lip	8	30%
Upper lip	11	41%
Nose	8	30%
Scalp	8	30%
Temple	8	30%

Number responding = 27 / percent responding = 19% of total respondents (143).
Note: This was a multiple choice question.

Page 45. III. Associated Symptoms (page 3 of 4) cont.: Numbness
2. Other locations of the body with numbness:

Other locations	Responses (25)	% of Number responding
Arm	5	38%
Back	4	31%
Chest	0	0%
Leg	10	77%
Neck	3	23%
Side	2	15%
Stomach	0	0%
Other (Specify) **Feet**	1	8%

Number responding = 13 / percent responding = 9% of total respondents (143).
Note: This was a multiple choice question.

Page 45. III. Associated Symptoms (page 3 of 4) cont.: Numbness

3. Comments for Numbness:

Responses (12)
I have one spot on my upper right leg feels numb.
When I was a child before i got sick that sensation only happened when I stood on a part of my body and did not let the blood circulate but now it just happens and it's like I don't have those parts of my body I don't feel them.
This may be more tingling than numbness, or maybe both when it happens.
comes and goes
The numbness is constant for me.
Left side of body, loss of pain sensation due to PRS affecting right side of brain.
small area on knee
My forhead and muscles for my eyebrow are numb due to the surgeries I have had. After 22 yrs. I am able to move my eyebrow a little.
May not be PRS related at all, I also have Chiari Malformation Type I.
I'm on Topomax, so its hard to say whether or not these are from the PRS, or that... also, I have Lupus, so whose to say what causes what..???
I think most of the numbness came because of the surgeries.
I am not sure excatly where the numbness occurs.

Number responding = 12 / percent responding = 8% of total respondents (143).

Page 46. III. Associated Symptoms (page 3 of 4) cont.: Problems with balance

1. Has any therapy been tried to improve this problem?

Linked	Responses (26)	% of Number responding
Yes	5	19%
No	21	81%

Number responding = 26 / percent responding = 18% of total respondents (143).

Note: If "Yes" was selected at the above question 46. III. 1. "Has any therapy been tried to improve this problem" then the following section (Page 47) would apply.

Page 47. III. Associated Symptoms (page 3 of 4) cont.: Problems with balance - therapy

1. Please describe the therapy:

Responses (3)
"Retraining the brain" exercises prescribed by physio for balance and falls prevention e.g. - standing tandem stance (heel to toe, feet straight) and holding for 2 - 3 minutes, progressing to doing this in bare feet on a spongy mat and with eyes closed - walking a straight line heel to toe - progressing to doing this moving head up and down or side to side. There are many exercises along these lines.
Physical and Occupational
WORKING ON CORE STRENGTHENING

Number responding = 3 / percent responding = 2% of total respondents (143).

Page 47. III. Associated Symptoms (page 3 of 4) cont.: Problems with balance - therapy

2. Please describe how successful it was:

Responses (3)
I have had great success with this therapy and it did stop the falls.
Physical Therapy very successful - affected Leg very strong. Occupational Therapy is not so good. Effected arm/hand has very limited mobility.
NOT VERY

Number responding = 3 / percent responding = 2% of total respondents (143).

Page 47. III. Associated Symptoms (page 3 of 4) cont.: Problems with balance - therapy
3. Comments concerning problems with therapy:

Responses (2)
Kept getting weak with pneumonia so would have to stop surgery.
Once you stop the exercises the balance problems do eventually return - need to keep them up on a regular basis.

Number responding = 2 / percent responding = 1% of total respondents (143).

Page 46. III. Associated Symptoms (page 3 of 4) cont.: Problems with balance
2. Comments concerning problems with balance:

Responses (16)
PHYSICAL THERAPY IS ONGOING
Only now and again very mild. Had a big problem after major micro vascular surgery. Took a few weeks to get back to normal with the balance.
It's not that bad. It's like I have anemia and my body sudenly relaxes and I have no control of it but those episodes are very rare and short.
My guess is that it is related to fatigue and headache. Sometimes I feel like my brain is misfiring (it's hard to explain) and I feel like I am going to tip over or am falling.
Sometimes head feels like it's not connected to neck.
I am constantly bumping into walls and doors.
Difficulty with moving stairs in shopping centers. Takes longer to regain balance after bending down or turning around.
Has physical and Occupation Therapy at school 3 times per week and at a Children's Hospital 1 time per week.
Did have some issues earlier where I fell over a lot but these seem to have gone now. Do have balance issues when I have a migraine.
This happens if I get tired and run down - starts with running into walls and doors progresses to tripping and then finally falling over.
Most likely due to Chiari Malformation. Trouble standing steady on both feet, especially with eyes closed.
I haven't had therapy for this.
Nothing major that I'm concerned with. Its more occasional at this time.
My daughter has been diagnosed with vertigo and she had one very severe bout with it. It impacted her balance.
I didn't know that Rombergs might be responsible for my slight problems with balance, I thought that it was due to anti-convulsives.
After the orginal surgery.

Number responding = 16 / percent responding = 11% of total respondents (143).

Page 48. III. Associated Symptoms of Rombergs (page 4 of 4)

1. Please select all that apply (page 4):

Symptoms	Responses (391)	% of Number responding
Scalp affected	55	46%
Sensitivity to extreme temperatures	42	35%
Skin, absence of pores	16	13%
Skin is discolored	80	67%
Stress	35	29%
Teeth affected	66	55%
Thin skin	78	66%
Other medical conditions not listed above	19	16%

Number responding = 119 / percent responding = 83% of total respondents (143).
Note: This was a multiple choice question.

Page 49. III. Associated Symptoms (page 4 of 4) cont.: Scalp affected

1. Please select how the Scalp is affected:

Scalp is Affected	Responses (155)	% of Number responding
En coup de sabre (indentation)	32	59%
Indentations	36	67%
Tenderness (Occipital neuralgia)	20	37%
Thin skin	33	61%
Tissue loss	28	52%
Other (Specify)	6	11%

Number responding = 54 / percent responding = 38% of total respondents (143).
Note: This was a multiple choice question.

Page 49. III. Associated Symptoms (page 4 of 4) cont.: Scalp affected
1. Please select how the Scalp is affected:
Other (Specify)

% of Total Responses	Responses (6)	Other - Scalp Affected
0.7%	1	skin disorders of the scalp
0.7%	1	hair loss
0.7%	1	surgery
0.7%	1	hair loss along the entire line of it
0.7%	1	bald spot
0.7%	1	no hair on part of scalp

Page 49. III. Associated Symptoms (page 4 of 4) cont.: Scalp affected
2. Comments for Scalp affected:

Responses (17)
I get rashes on my scalp that are full of pus and very scaly. Sometimes it will spread and go down my neck and to my eye lids and forehead. I have a very flaky and itchy scalp too. The only time I get this is when i have been under a lot of stress or on rare occasions get my hair colored or use a different shampoo. Also some medications can cause it too.
There is an indentation on the right side of my scalp.
3 very deep 'dents' in scalp-90% of top/front scalp
It's only thin skin and less hair than on the parts that are not affected.
My MRI noted "Thinning of the scalp."
The scalp was affected from the surgery. I had a section of scalp removed and implants in the lower right side of my face. 90% of the hair grew back but there is a bald spot on the right side. This bald spot is covered by hair but is tender to the touch and the skin is very thin and discolored.
Even a slight bump on something of the indentation will cause me to nearly pass out. It's very sensitive and I am on automatic to protect that area.
Terribly, loss of alot of hair, I have about 8 bald patches to about 12 patches. All my hair has thinned out.
The scalp area has been affected only since it has restarted again a few years ago and has become a major problem for me. It is spreading faster and further than it ever did.
a few little bald spots
indentions and hair loss
just very sensitive
I had a bald spot in the middle of my head when Romberg's first happened. Also on my left side of my head it is slightly indented. The last surgery I had in 2002 he fixed my bald spot by cutting and stitching it back together.
bald spot right side with odd skin there looks like a burn
Hurts to comb hair. Very sensitive to touch.
There is a long strip of the scalp that has severe indentation or soft spot, and no hair grows in this area.
En coup de sabre is 3 in. long on right side.

Number responding = 17 / percent responding = 12% of total respondents (143).

Page 50. III. Associated Symptoms (page 4 of 4) cont.: Sensitivity to temperatures

1. Please describe Sensitivity to extreme temperatures:

Describe sensitivity	Responses (65)	% of Number responding
Hands sensitive to cold	26	76%
Hands sensitive to heat	4	12%
Feet sensitive to cold	22	65%
Feet sensitive to heat	4	12%
Other (Specify)	9	26%

Number responding = 34 / percent responding = 24% of total respondents (143).
Note: This was a multiple choice question.

Page 50. III. Associated Symptoms (page 4 of 4) cont.: Sensitivity to temperatures

1. Please describe Sensitivity to extreme temperatures:

Other (Specify)

% of Total Responses	Responses (6)	Other - Sensitivity To Temperatures
1.5%	1	upper lip numb during cold temp
1.5%	1	scalp sensitive to cold
1.5%	1	face
1.5%	1	everything
1.5%	1	have had Raynaud's Syndrome since a teen
1.5%	1	arthritis
1.5%	1	right side of face
1.5%	1	not sure
1.5%	1	depending on the weather, depends on how she feels

Page 50. III. Associated Symptoms (page 4 of 4) cont.: Sensitivity to temperatures

2. Comments for Sensitivity to temperatures:

Responses (17)
Left upper lip numb during cold temp or easily sunburns.
Under extreme stress my blood vessels in my hands and feet will throb and shrink up. My skin will get tight and bluish or purplish pink in color.
When it's cold my hands and legs are numb and the skin is at first yellow and after that, like I have bruises and at heat especiallly in the sun, the skin burns very fast and I lose after that skin for 3 - 5 times depends on the gravity .... it's with small balloons of water under the skin 2 - 5mm.
I have Raynaud's Syndrome.
Seems like I don't tolerate either extreme as well as others do.
My face hurts when it is cold.
Feet sensitivity may be related to having tendon surgery, not to PRS.
Face is sensitive to cold.
When my PRS is active I have sensitivity to the cold. Normally I don't have a problem as I am naturally very insulated! I really notice it when swimming etc., everyone else says the water is fine but it is freezing to me and I just can't stay in.
Cannot tolerate either extreme cold or extreme heat.
Again, most likely due to Arnold Chiari. My hands seem slow to respond to hot temperatures, eg. taking food out of the microwave, I won't be able to tell if it's hot or just warm, I'll have to use the back of my hand.
I thought I was just cold blooded?? Also, Lupus people have Raynaud's Syndrome... not sure if that is the cause or not, or if they are related...
Cold sensitive due to the titanium plates.
I believe the sensitivity is only in the affected areas, that does not include hands or feet.
Not sure, face gets red from heat or cold.
I hate the cold!!!!!
I have Raynaud's which started at six or seven.

Number responding = 17 / percent responding = 12% of total respondents (143).

Page 51. III. Associated Symptoms (page 4 of 4) cont.: Skin is discolored
1. Please select the location of discoloration on the head:

Affected	Responses (172)	% of Number responding
Cheek	41	54%
Chin	26	34%
Ear	4	5%
Eye lid	14	18%
Forehead	24	32%
Lower lip	11	14%
Upper lip	18	24%
Nose	9	12%
Scalp	10	13%
Temple	15	20%

Number responding = 76 / percent responding = 53% of total respondents (143).
Note: This was a multiple choice question.

Page 51. III. Associated Symptoms (page 4 of 4) cont.: Skin is discolored
2. Other locations of the body with discoloration:

Other locations	Responses (54)	% of Number responding
Arm	4	12%
Back	12	35%
Chest	6	18%
Leg	8	24%
Neck	14	41%
Side	2	6%
Stomach	3	9%
Other (Specify)	5	15%

Number responding = 34 / percent responding = 24% of total respondents (143).
Note: This was a multiple choice question.

Page 51. III. Associated Symptoms (page 4 of 4) cont.: Skin is discolored
2. Other locations of the body with discoloration:
Other (Specify)

% of Total Responses	Responses (5)	Other - Locations With Discoloration
1.9%	1	belly
3.7%	2	under eye
1.9%	1	not sure
1.9%	1	breasts

Page 51. III. Associated Symptoms (page 4 of 4) cont.: Skin is discolored
3. Please rate the severity of this discoloration:

Rating	Responses (77)	% of Number responding
1 (very mild)	10	13%
2 (mild)	23	30%
3 (moderate)	31	40%
4 (severe)	11	14%
5 (very severe)	2	3%

Number responding = 77 / percent responding = 54% of total respondents (143).

Page 51. III. Associated Symptoms (page 4 of 4) cont.: Skin is discolored
4. Comments for Discoloration:

Responses (24)
Vitiligo was first known sign of PRS. Vitiligo then turned a full 360 degrees into dark pigmentation.
Discoloration very noticable in early years - on lower chin - white patch began. Lip noticably bluish tinges. Then area on cheek became pale in color. All much improved since reconstructive surgeries.
I guess not enough blood goes through those areas.
The area that has lost tissue has become discolored... mostly around the right side of my neck.
Medium brown colour, shiny.
Has changed colors... started of pink, then red, then blue/white and then tan. Now a darker brown/gray color.
Dark areas can look bruised at times, very thin skin so blood vessels can be seen.
No discoloration on head.
It's like the blood is missing from that area.
Easily covered with bare minerals makeup.
Mostly red blemishes on forhead on left side and on check but can cover up with make up.
Bluish tint where there is a lack of tissue.
It's worst on my forehead and neck.
The eye discoloration is under the eye (looks like a black eye) not on eyelid.
Only because where there is little muscle/fat the skin is more translucent - can see the blue veins underneath.
Cheek gets really red if outside in the cold!! On the plus side - NO ZITS on affected side!!! Woo hoo!!
I would say severe... dark brown in color. Doctors would probably say moderate.
I think the skin on my right side is getting thinner as the veins are showing through on that side and it is much more lined of course. Under my eye is noticeably darker now and I have to use cover-up under my make-up.
Looks like suffering from mild vitiligo.
"Chin" above is underneath her chin. It seems to vary with available light and with the day for some reason.
There are areas of her face that have no pigment.
The discoloration only started a few years ago (I am 29 now, diagnosed when 8). Just when you think you've had it all and the disease has come to a stop.....
Discoloration on upper right and lower right side.
This came very late, in my second round of symptoms in my late 40's.

Number responding = 24 / percent responding = 17% of total respondents (143).

Page 52. III. Associated Symptoms (page 4 of 4) cont.: Stress
1. Please select how stress affects the person with Rombergs:

Affected	Responses (137)	% of Number responding
Anxiety	27	77%
Daily life struggles	19	54%
Depression	21	60%
Difficulty in relationships	14	40%
Difficulty with job opportunities	10	29%
Panic attacks	12	34%
Self esteem issues	29	83%
Other (Specify)	5	14%

Number responding = 35 / percent responding = 24% of total respondents (143).
Note: This was a multiple choice question.

Page 52. III. Associated Symptoms (page 4 of 4) cont.: Stress
1. Please select how stress affects the person with Rombergs:
Other (Specify)

% of Total Responses	Responses (5)	Other - How Stress Affects
0.7%	1	terribly effects me
0.7%	1	I'm only speaking for myself here..
0.7%	1	extreme fatigue
0.7%	1	frustration
0.7%	1	not sure

Page 52. III. Associated Symptoms (page 4 of 4) cont.: Stress
2. Do you feel stress has a negative impact on Rombergs?

Affected	Responses (33)	% of Number responding
Yes	30	91%
No	3	9%

Number responding = 33 / percent responding = 23% of total respondents (143).

Page 52. III. Associated Symptoms (page 4 of 4) cont.: Stress
3. Comments for Stress:

Responses (14)
I notice when I am stressed, I get more headaches, migraines, and eye problems.
I feel stress can bring on anything.
Everything
I lose my temper quickly sometimes it scares me I am afraid I will do something I will regret later.... my face it just burns in that moment and I have a smarting pain sometimes it changes it's color and it's red and my veins..... like they will blowup.
I wonder if my stress has escalated the process of Rombergs (the atrophy). I am a full- time student, work full-time, and juggle life in general.... so I wonder if that caused the progression so quickly.
I think the frustration of there not being adequate medical knowledge and treatment is a major factor in Romberg related stress. The lack of knowledge and interest by doctors is astonishing and frustrating. I go into a panic when I have to see a doctor about my PRS symptoms because I am so often told I am not experiencing what I say. This even though the doctors have never even heard of PRS, so how could they know? The only doctor I have ever seen who knew anything about PRS was the plastic surgeon who diagnosed me. Some of the worst are neurologists. Some of the best are physician assistants — they at least are not dismissive and seem to want to help and find out more.
I get stressed whenever I notice my Rombergs is progressing. Other than that, life pretty much goes on and I don't think about it. Also, I was under a lot of pressure at work last year which seemed to speed up the progression.
Seems to 'burn' when life gets tough!
I don't get stressed from Rombergs but have the normal stresses of everyday life... my Rombergs had been quiet for nearly six months and then some major family drama involving a major car accident and my husband and myself separating it is now active in a big way with my face tingling and itching constantly... though it is reducing as things get sorted.
Stress has a HUGE affect in this case.
I'm scared for me and my children. I was never so anxious before as I am every day now. In fact, I didn't know what anxiety really was until this came into our lives. How do you not despair about what is or might be happening to you or your child when you see the changes?
I feel that it really gets my PRS going, but then again, it really affects my Lupus too!
My daughter needs to find a niche with as little stress as possible. She is taking medication for depression at this time as well, to my chagrin.
When she becomes over tired or stressed she gets infections in her face. She is on antibiotics on a daily basis to prevent these severe and painful outbreaks.

Number responding = 14 / percent responding = 10% of total respondents (143).

Page 53. III. Associated Symptoms (page 4 of 4) cont.: Teeth affected

1. Please select how the Teeth are affected:

Affected	Responses (218)	% of Number responding
Braces	14	21%
Delayed eruption	17	26%
Loose	13	20%
Loss of	21	32%
Misaligned	39	59%
Missing	14	21%
No permanent teeth	8	12%
Overbite	12	18%
Overcrowding	16	24%
Receding gums	23	35%
Resorbed roots	14	21%
Root decay	16	24%
Other (Specify)	11	17%

Number responding = 66 / percent responding = 46% of total respondents (143).
Note: This was a multiple choice question.

Page 53. III. Associated Symptoms (page 4 of 4) cont.: Teeth affected
1. Please select how the Teeth are affected:
Other (Specify)

% of Total Responses	Responses (11)	Other - How Teeth Affected
0.5%	1	bone loss
0.5%	1	short root at 3 teeth
0.5%	1	short roots
0.5%	1	dwarf wisdom teeth
0.5%	1	smaller on PR side
0.5%	1	grind teeth
0.5%	1	missing bottom wisdom tooth
0.5%	1	nerve atrophy
0.5%	1	had braces, but no longer
0.5%	1	open bite
0.5%	1	not sure

Page 53. III. Associated Symptoms (page 4 of 4) cont.: Teeth affected

2. Comments for Teeth affected:

Responses (22)
I have only 7 teeth on the bottom gums of my mouth several at the top and some have shifted.
They are sensitive, sometimes I have pains but there is nothing wrong no cavities or damages and sometimes I feel like they are moving and they'll fall.
single front tooth
I have lost 2 upper teeth on my left side.
On the affected side 3 teeth did not develop properly.
Missing wisdom teeth on that side.
I lost several teeth on the affected side and will also need a bone graft due to continuing bone loss.
Braces corrected bite nicely--- Romberg's folks aren't the only ones with braces-- just remember that.
The majority of my teeth have problems with the root area and bone loss.
Have lost most of the top left side teeth.
ongoing problems now
We are not sure if it from her disease, but her teeth are a year behind in growth and she has a 100% overbite the dentist said.
Having full mouth xrays taken at end of month. Seems cannot find some of her adult teeth. Follow up with dentist.
One tooth was effected due to PR. Teeth on left side are smaller than teeth on right side.
Missing one lower adult tooth that never developed on the Romberg's side.
Palate and tongue on left side are smaller.
Probably has nothing to do with Rombergs. My brother who is not affected has the same.
They have been more misaligned over the years.... I keep looking in the mirror to keep track of them... luckily, not the front ones, just the ones on the left/affected side...
open bite on affected side
bone loss and root decay, no top teeth
Braces when she was first diagnosed and then again in teenage years. Some teeth removed because of overbite.
Her teeth are awful, very tiny.

Number responding = 22 / percent responding = 15% of total respondents (143).

Page 54. III. Associated Symptoms (page 4 of 4) cont.: Thin skin

1. Please select the location of Thin skin:

Affected	Responses (274)	% of Number responding
Cheek	46	59%
Chin	37	47%
Ear	7	9%
Eye lid	24	31%
Forehead	40	51%
Lower lip	17	22%
Upper lip	28	36%
Nose	23	29%
Scalp	21	27%
Temple	25	32%
Other (Specify)	6	8%

Number responding = 78 / percent responding = 55% of total respondents (143).
Note: This was a multiple choice question.

Page 54. III. Associated Symptoms (page 4 of 4) cont.: Thin skin

1. Please select the location of Thin skin:

Other (Specify)

% of Total Responses	Responses (6)	Other - Location Thin Skin
0.4%	1	stomach, foot, leg, back, arm
0.7%	2	neck
0.4%	1	back
0.4%	1	AROUND THE EYE
0.4%	1	under right eye

Page 54. III. Associated Symptoms (page 4 of 4) cont.: Thin skin
2. Comments for Thin skin:

Responses (10)
Much more noticable in early years. Same has been helped by improved blood supply following reconstructive surgeries.
I have none.
Although it's thin, I don't have acne on that part, but i do have like small points of fat and it sweats.
Very thin shiny tight skin, medium brown colour. No fatty tissue underneath.
Chin has most atrophy, followed by nose and lip.
The skin is thinnest on my forehead.
Can see veins under the skin.
Weird.... thin- can see veins.
Discolorations and tender areas.
Lower lip and upper lip on the right side are wrinkled.

Number responding = 10 / percent responding = 7% of total respondents (143).

Page 55. III. Associated Symptoms (page 4 of 4) cont.:
Other medical conditions
1. Please describe other medical conditions not listed above:

Responses (16)
arthritis, lop sided body
Horners Syndrome where the pupil of my left eye does not dilate.
I have been recently refered to a Rheumatologist because I test positive for Anti ds DNA. I have also been suffering from total body pain and illness. I get Pyelonephritis with kidney stone along with SEPSIS if an infection is involved. I suffered from endometriosis for years and was diagnosed with secondhand infertility until 2 years ago when I got pregnant with my 2nd child. Also, I am hypersensitive to medications. I will get an allergic reaction to almost anything.
I have a cyst on one of my kidneys, 8 mm I had one on my up eyelash this one I operated and I have a small one on my right knee........... I also have cyst on my ovary I have problems with my stomach and the intestines. I have weight problems and I have negs on my skin. I have problems with learning. I can concentrate I understand but I don't assimilate the information .... and sometimes I just can't remember the action with a few moments before.
asthma allergies hypertension
fibromyalgia, chronic fatigue syndrome
BRAIN!!! loss of motor neurones on right side spastic paralysis on left side of body
Bipolar disorder, I began with depression following my reconstructive surgery.
Back pains, I developed a mild case of Scoliosis from the muscle atrophy in my back.
Duane's Syndrome, one eye cannot look left. May or may not be related to Rombergs. Possible Raynaud's Syndrome also. Mother has this too.
During 2nd pregnancy, back, hip, knee and ankle problems that got so severe I was unable to walk and had to use a wheelchair for 1 month before and 1 month after childbirth. Since childbirth, my mobility has gradually been returning to normal. These problems were never formally diagnosed but could have been rheumatoid issues or SPD. Unclear whether related to PRS on not - I did not experience them during my 1st pregnancy, only during the 2nd one.
acid reflux, asthma, migraines, seizures, arthritis, loose joints,
Learning challenges, particularly with vocabulary and expressive language. Vision difficulty, as her left eye is deeper and lower than her right eye. This causes double vision, and probably also contributes to the migraine headaches. She is not a 'reader', and all of the above contribute to this. Her family is more into reading, so this is an unusual trait.
No sure which of her problems are related.
Progressive hearing loss, inflammation of cochlear nerve and depression are being treated.
asthma, sinus problems, slight hearing lose, eye movement problems, seizures, migraines, behavorial, arthritis

Number responding = 16 / percent responding = 11% of total respondents (143).

Page 55. III. Associated Symptoms (page 4 of 4) cont.:
Other medical conditions
2. Comments for Other medical conditions:

Responses (6)
not sure related problems
low blood sugar high cholesterol (family history)
chronic pain at the donor site in my chest
Dad has Ankylosing Spondylitis
none
Don't know if these conditions are related to Rombergs but my small left side is the side affected by arthritis.

Number responding = 6 / percent responding = 4% of total respondents (143).

Page 56. IV. Triggers for Rombergs (page 1 of 2)

1. What possibly could have triggered Rombergs? (page 1):

Triggers	Responses (155)	% of Number responding
Affected person has an autoimmune disorder	29	33%
Affected person was breast fed	23	26%
Dental work or tooth extractions prior to onset	21	24%
Experienced head trauma of any kind prior to onset	52	60%
Live near a polluted area or any factories prior to onset	11	13%
Lyme disease	5	6%
Miscarriage prior to onset	1	1%
Serious infection of any kind prior to onset	13	15%

Number responding = 87 / percent responding = 61% of total respondents (143).
Note: This was a multiple choice question.

Page 57. IV. Triggers (page 1 of 2) cont.: Autoimmune disorders

1. Please select any Autoimmune disorders that the affected person has:

Autoimmune disorders	Responses (48)	% of Number responding
Addison's disease (adrenal)	0	0%
Ankylosing spondylitis	0	0%
Autoimmune Thyroid disease	5	18%
Arthritis	6	21%
Celiac disease	0	0%
Chiari Malformation	1	4%
Chronic Fatigue Syndrome	2	7%
Crohn's disease	0	0%
Fibromyalgia	2	7%
Goodpasture's Syndrome (lungs, kidneys)	0	0%
Graves' disease (thyroid)	0	0%
Guillain-Barre Syndrome (nervous system)	0	0%
Hashimoto's thyroiditis	3	11%
Hughes Syndrome (antiphospholipid)	0	0%
Inflammatory bowel disease	2	7%
Lupus (SLE)	1	4%
Mixed Corrective Tissue disease	0	0%
Multiple Sclerosis (MS)	0	0%
Polymyalgia Rheumatica (large muscle groups)	1	4%
Raynauds Phenomenon	3	11%
Scleroderma (skin, intestine, less commonly lung)	0	0%
Scleroderma, linear	8	29%
Scleroderma, localized	6	21%
Sjogren's Syndrome	0	0%
Systemic sclerosis	0	0%
Temporal Arteritis / Giant Cell Arteritis (arteries of the head and neck)	0	0%

Continued on the next page.

Autoimmune disorders	Responses (48)	% of Number responding
Thyroid problems	4	🟥 14%
Type 1 Diabetes Mellitus	0	0%
Ulcerative colitis	0	0%
Vitiligo	4	🟦 14%
Other (Specify)	5	🟥 18%

Number responding = 28 / percent responding = 20% of total respondents (143).
Note: This was a multiple choice question.

Page 57. IV. Triggers (page 1 of 2) cont.: Autoimmune disorders
1. Please select any Autoimmune disorders that the affected person has:
Other (Specify)

% of Total Responses	Responses (5)	Other - Select Any Autoimmune Disorders
2%	1	undifferentiated Connective Tissue
2%	1	autoimmune
2%	1	psoriasis
2%	1	just have PRS
2%	1	Lichen Planus

Page 57. IV. Triggers (page 1 of 2) cont.: Autoimmune disorders
2. Comments for Autoimmune disorders:

Responses (11)
Thyroid problems run in family in the women. My mother has had both underactive and overactive and a goiter.
Not sure if this applies here, but in case I forget..... I was 13 when I first became ill with Lupus.... had been out collecting sap with an uncle and cousins and came down with an awful case of strep throat... that turned into chest pains, etc.. eventually went to the school nurse who took my temp (after strep cleared up) she called my mom to take me to hospital, who in turn did an X-ray of my heart which was enlarged to 2X normal size - years later did they finally diagnose Lupus. This was all around a year or so before first signs of PRS showed up - the discoloration near chin - in fact, I first asked my Lupus Dr. about it. He said he didn't know what it was.... hmmm??
Eczema? Not sure if it's autoimmune or not.
Ugh
I have had Lichen Planus for over 25 / 30 years. I am asymptomatic, but since it is a pre-cancerous condition, as recommended by my dentist, I now see a specialist at the University of Pennsylvania every 6 months just to keep a check.
I believe that PRS is an autoimmune disorder.
I have seen an autoimmue specialist and she has done extensive testing for autoimminue disorders. The results have shown that I do test positive for autoimmune disorders. I have Lucopenia (a chronic low white count) and other autoimmue disorders.
My blood tests are also positive for scleroderma, and I had Herpes Zoster just before the onset of the Romberg's symptoms.
alopecia areata, eczema, sinus canal problems
none
very demoralizing

Number responding = 11 / percent responding = 8% of total respondents (143).

Page 58. IV. Triggers (page 1 of 2) cont.: Breast fed

1. How long breast fed:

Count	Responses (21)
1	2 years
1	a few months
1	about 9 months
3	18 months
2	1 year
1	4 months
1	11 months
6	6 months
2	2 months
2	3 months
1	7 months

Number responding = 21 / percent responding = 15% of total respondents (143).

Page 58. IV. Triggers (page 1 of 2) cont.: Breast fed

2. Comments for Breast fed:

Responses (7)
I am the youngest of eight children. My mom was in pretty poor health by the time I was conceived / born.
Did fine, Mother went out of town for 10 days, returned and try to resume breast feeding but it was unsuccessful at that point. But otherwise the breastfeeding was fine.
Surely that's not a cause for Rombergs?! Its good for you!
Mother went back to work and bottle fed breast milk to 1 year or so.
Baby led weaning at 18 months.
Was also formula fed for the first year, as I worked and had to supplement.
Rombergs can start at any age so I cannot believe breast feeding has anything to do with triggering Rombergs. Billions of babies are breast fed but do not get Rombergs. In fact breast milk is far more nutritious & has many immunisation qualities which would fight infection & diseases. I would think babies who were not breast fed would be more prone to Rombergs.

Number responding = 7 / percent responding = 5% of total respondents (143).

Page 59. IV. Triggers (page 1 of 2) cont.: Dental work
1. Comments for Dental work:

Responses (19)
Silver fillings when a teenager.
At 16, my son had his upper right molar removed. During the operation the dentist broke his tool and the tooth was catapulted into the upper jaw. A piece of bone from the jaw of approximately 2 inches in diameter, was cut from the jaw, in order to remove the tooth. The healing process was very long and there have been sinus problems ever since. Somehow I think that this operation was the trigger of the PRS, especially as the eye began to recede about 3 month later, resulting in an enophthalmus and atrophy of the eye and right side of the face.
i HAD TO HAVE A TOOTH EXTRACTED AT THE AGE OF 6 (ON THE SIDE THAT EVENTUALLY HOSTED ROMBERGS)
From looking at photos of my teen years, it looks as though I had some atrophy on the left side of my face. However, after I had my wisdom teeth removed when I was approximately 24 was when I first noticed that something was wrong with my face/cheek.
Braces for 2 years and dark spot and jaw problem start to show. Maybe he have it before but we never see that on him.
Dentist used me as a personal lab rat. They didn't know what would work and what wouldn't. I have been through a medical malpractice suit due to a dentist not acknowledging that I have PRS.
Had a difficult wisdom tooth extraction.
I think I was 8 when I had a molar that had to be taken out. I was afraid of the dentist and did not complain for a long time. Besides that, I do not experience a lot of pain when it is about my teeth, so I could hold on quite a long time.
He had teeth pulled before onset.
In 2005/2006, I had extensive dental work done---upper bridge, lower bridge, partial plate. I did have 2 lower teeth extracted, but I think those extractions were in early December 2006 and I noticed the atrophy of my right cheek in early 2007.
amalgam dental filling
Poor baby teeth, many extractions - particularly a large molar on what became my affected side.
Braces for 18 months - 2000- 2001
Braces as a child. Also broken front tooth as a result of falling over and losing half of it as a child.
Was missing 1 wisdom tooth! Not sure if on the PRS side or not. Gums have definitely receeded, mostly on PRS side.
Teeth are a mess. Found permanent teeth laying in gums, pieces of bone fragments laying in gums, no permanent teeth left side, teeth very tiny, teeth always feels loose, never eats on left side, needs braces, teeth are all shifting to right side. Gums are eating away, overbite.
Braces at 12 yrs, for 1 year.
The only thing I can say is I have had a wisdom tooth pulled before my diagnosis and other routine dental procedures.
She had a crown put on at age 4. That very day her lip drooped.

Number responding = 19 / percent responding = 13% of total respondents (143).

Page 60. IV. Triggers (page 1 of 2) cont.: Experienced head trauma
1. Please rate this trauma as:

Rating	Responses (49)	% of Number responding
Major trauma	12	24%
Minor trauma	37	76%

Number responding = 49 / percent responding = 34% of total respondents (143).

Page 60. IV. Triggers (page 1 of 2) cont.: Experienced head trauma
2. Describe the location of this trauma:

Count	Responses (46)
1	Forehead, above right (affected) eye.
1	I had a garage door fall on my forehead.
1	Right side face
1	Huge head bang - knocked unconscious in road accident.
1	I got kicked in the left jaw and chin with a big man's foot while i was taking swimming lessons while living in Louisianna. The same week I got pushed on a log in the water while I was playing with a boy. It tore up my right back, I was bleeding from it. I was 8 years old when this happened.
1	Neck
1	I cut my forehead open on the left side at the hair line, approximately intersected at the part line.
1	Fell out of a car and smashed my head on the curb. Car moving slowly, but still remember the accident. Happened when I was very young, maybe 3 or 4 years of age.
1	When I was approximately 2 years old, I was standing on the toilet seat (my sister was brushing my hair) and I wiggled away from her and fell into the bathtub, hitting the side of my head, (I don't know which side).
1	Hit with a baseball bat on my forehead at approx age 7. Also, was propelled onto a propane tank on which I hit the back of my head and also hit it again as I fell onto the cement base of the tank. (Brothers!!!) Approx age 7. Both required trips to the ER. (My poor Mom)
1	Chin. Also fell on face, but we don't remember what side.
1	I was in a car accident where the car was rolled and I hit my head about 8 times as the car rolled.
1	Forehead broken
1	Got hit incredibly hard on my head with a moving wooden swing. This incident took place about one year prior to start of PRS.
1	Experience head trauma as a child. Playground accident where I fell to the ground and hit my head.
1	Stitches in upper lip due to injury of being hit with snow shovel.
1	Center of forehead
1	I fell on cement steps and hit my nose. The symptoms began to present after the fall.
1	Went over handlebars of my bike and landed on my face on the pavement. Also fell off a picnic table and landed on my face. Also fell off the monkey bars and got a concussion (I was a pretty active/accident prone kid!).

Continued on the next page.

Count	Page 60. IV. 2. Responses (continued)
1	Middle top of forehead
3	Forehead
1	I was in two minor traffic accidents. My car was rear-ended two different times, about a year or two apart in the late 90's. I experienced some neck pain on my right side.
1	At age of 2 he got a trauma from a wood in the same area, where he got the coup de sabre.
1	Hit head against sidewalk - loss consciousness for a very short time, mild concussion.
1	Wore the helmet for approximately 8 months from age 12 - 18 months.
1	Small scratch from pet dog right at the spot where it started.
2	I was delivered by forceps and badly bruised.
1	Ice skating rink
1	Top of head
1	1. Top of head, possibly face 2. Chin
1	Fall on head on age 4.
1	Fell and had a hard knock on the head.
1	I was ejected from an auto when I was nineteen. I suffered an intercraniocerebral trauma. This left me with diplopia and poor strength and miscoordination of my left upper extremity. When I was 52 I had strabismus surgery on my eyes. One eye had to be rotated and the other eye had to be raised. Condition was not 100 percent corrected, but close. When I was ejected from the car I must has landed on my head because I had a cut about 25 mm. long on the top/rear of my head. In the area of the normal cowlick a person has.
1	She bumped her head on the floor, falling from Mom's hip down to the floor.
1	Fell, hit forehead on brick steps, had stitches.
1	Right jawline
4	Head
1	Face
1	Struck forehead when she fell and needed stitches.
1	This blunt trauma occured after the orginal diagnosis of the PRS, but it triggered it out of remission and resulted in the seizures and lesions.

Number responding = 46 / percent responding = 32% of total respondents (143).

Page 60. IV. Triggers (page 1 of 2) cont.: Experienced head trauma

3. Comments for Experienced head trauma:

Responses (27)
Three layers of stiches for a deep laceration that went down to skull.
Ran into a tree while snow tubing at 12 years of age.
Got hit while playing volleyball, could not move neck for three days, went to hospital with sprained muscle.
Unsure, but typical childhood trauma. Fell about 10 feet from a tree at age 7.
I may have had a head trauma when I was born, parents still not talk about it. Had an inclination from an Uncle something happened.
When I was 5 years old I was running with my head down and ran full force into a corner of a wall. It knocked me down and cut my head open where I needed approximately 5 stitches.
I'm told I use to rock back and forth and bang my head against a cement wall and other surfaces when I was a toddler. I suspect this was due to severe neglect as an infant/toddler. But, perhaps, it was a response to pain with the onset of Rombergs? Since I was young I don't know if the onset or active part of the Rombergs is a painful process. I was 3 when the Rombergs began and from looking at pictures it looks like a majority of the atrophy in the forehead happened rapidly, though the indentation in the nose happened later. My parents are of little help with information.
See above
I am not sure if this caused it to happen, but did not have any problems until about a year after this accident.
Fell down stairs at an early age and hit head.
Fell down the stairs and hit the wall. Both eyes were black and blue.
Forehead broken (yes it sounds strange but it is true) after I had fallen from a hill. This was in 1972 the year that PRS started. Back then the technique wasn't that good (I lived on a small Island in the Caribean) and the doctors were discussing what to do. After TWO WEEKS, having me complaining about severe headaches, they decided to operate me anyway. Time was running out. Everthing seemed to be ok after that. Later we thought the doctors may had triggered something causing my chin not to grow further. Now we know that isn't the case and that it is PRS.
Fell onto concrete floor and needed stitches on my forehead/
Fell of bed. Nothing seen in cscan or xrays.
Egg sized bump from being hit by a rock followed by a thin white line down this same area. Five years later thin indented line appeared vertically in same area and progressed through scalp to crown of head.
Slammed the boot of my car onto my head. Lock mechanism hit my head first. I nearly knocked myself out and there was a dent where the lock bit hit surrounded by swelling and bruising. I didn't even really notice the dent hadn't gone and had gotten bigger until around two months later.
Banged his head requiring stitches.
Downfall from table onto back of the head (aged 1). Downfall from horse onto (helmeted) back of the head (aged 3).
Might have been a coincidence.
Fell over on to my head. Blacked out for a few seconds, not concussed. Not sure if it was in 2002 or 2003.
Don't know if this is minor or major.
1. Had a difficult birth. Doctor used suction/vacuum 31 times in failed attempts to extract baby. (hospitals usually only allow its use 3 times) Baby had to be treated for headaches at birth. Was not breathing when born. 2. Fell on playground and hurt chin. Had to have stitches. Happened about 3 months prior to the first symptoms.
Continued on the next page.

Page 60. IV. 3. Responses (continued)
Sledding accident, hit a fence post knocked unconscious and had concussion.
She went to the doctor as a result of her fall and the doctor said she was fine. This was at about 10 months of age.
Head trauma was on upper right forehead. Parry Romberg is on left side of face. Trauma happened around the age of eight. Parry Romberg is visible retrospectively in photos starting after that age. Did not become significant enough for diagnosis until late teens.
Skating fall, not extreme, just enough to leave a bruise.
Not sure if its characterized as major or minor............. fell on my head when I was 5, running around a pool............. went to the hospital, lost vision for awhile and i think I had a concussion also.

Number responding = 27 / percent responding = 19% of total respondents (143).

Page 61. IV. Triggers (page 1 of 2) cont.: Live near a polluted area
1. Please comment on living near a polluted area:

Responses (11)
Grew up in an area that had a toxic waste dump approximately 7 miles away. (It has since closed down)
We moved from the country to Glens Falls, New York when before Katie was born. Prior to that my Mom had 8 kids in rural upstate. Katie and my brother were born after we moved. There were factories in our neighborhood, however my brother does not seem to be affected.
Live less then 1 mile from United Steel and DTE as well as several oil refineries. Lots of dust in the air.
Lower Manhattan during and after 9/11... was one of the first residents back.
Bronx, New York
Detroit area...motown. Pollution everywhere.
There was a mine where they dumped thousands of barrels of chemicals into and there is a big increase in cancer, which is currently being investigated by the government. This was located about 3 miles from where I lived and affected our water supply.
In the time.... the period when i was born was the accident with that nuclear explosion at Chernobyl........ I live at approximately 500 km from that place but I think that it could be a factor.
I grew up with factories down the road from me. You never thought about it growing up.
My grandparents and I lived in South East Texas. My grandfather was a welder and we moved around a little while he worked in the plants. We moved to Johnsons Bayou close to Christal Beach in Louisianna when I was 8 years old for 1 year. My grandfather worked in a Poggy Plant that stored fish. Later on I found out these fish had mercury in them. Also, some of these fish where brought home to eat. Also in South East Texas there have been plants that have caused different deformities in children, cancer, bronchitis, syndromes along with many other diseases caused by the pollutions they where sending out through the air and water. Some of these plants are under lawsuits.
I LIVED OUT IN THE COUNTRY WHERE THEY ROUTINELY SPRAYED WEEDS WITH CREOSOTE AND SPRAYED DDT FOR MOSQUITO CONTROL. PEOPLE BURNED THEIR GARBAGE BEHIND THEIR HOMES SO IT WAS RATHER UNSANITARY.

Number responding = 11 / percent responding = 8% of total respondents (143).

Page 62. IV. Triggers (page 1 of 2) cont.: Lyme disease
1. Please comment on Lyme disease:

Responses (5)
Prior to onset of PRS - maybe a few months after treated for Lyme.
Lyme disease was discoverd on age 7. After antibiotic: headaches milder and disappearance of tissue stopped.
Had a tick cut out of my right thigh in May 2004 -- had bulls eye, etc. Bulk of problems came after this, although photo shows spot on forehead in 2002.
Tested postivite for Lyme disease as an adult. Could have also been a false positive.
Never diagnosed with Lyme Disease, but bitten by tick which stayed in head 48 hours. Doctor said no Lyme disease-bearing ticks in Indiana so concluded I don't have it. Test inconclusive.

Number responding = 5 / percent responding = 3% of total respondents (143).

Page 63. IV. Triggers (page 1 of 2) cont.: Miscarriage prior

1. Please comment on miscarriage prior to Rombergs:

Responses (1)
During my 1st of 3 pregnancies, I had a miscarriage at around the 8th week. The results found that it was a partial Hydatidiform mole, an abnormal form of pregnancy.

Number responding = 1 / percent responding = 0.7% of total respondents (143).

Page 64. IV. Triggers (page 1 of 2) cont.: Serious infection

1. Please describe serious infections of any kind prior to onset:

Responses (12)
Staff infection as a child.............. thumb surgery needed, chicken pox, measles and mumps........... I think.
At age 4, had a sore/boil on my head where my brother hit me with a little metal cap gun that became severely infected!
Bad food poisoning a few times within a year.
At birth while still in hospital, I caught staph - I had it all over my head and face.
Strep throat, measles, chicken pox, blood poisoning.
Epstein-Barr-Virus
Had atypical pnuemonia in early 2002, 8 months after 9/11. Quite ill for awhile.
Some weeks before onset we observed 2 small centers of inflammation (like mosquito bites) on the right jaw. Inflammations endured for longer time (4 weeks or maybe longer).
My four children all had chicken pox for a month about six weeks before I hit my head. I had never had chicken pox and never got any symptoms and at the time I wasn't tested for it. I got some virus just after I injured my head and was exhausted all the time and had headaches and joint aches. I was tested for glandular fever and Ross River fever and other similar things but seem to have anything. It took six months before I started to get back to normal again!
Strep throat carrier (as far as we know).
Not an infection, but wanted to mention that I had a serious asthma attack at age 18 months, which my mother blames for the PRS. I was in intensive care and came close to not making it during that asthma episode.
Prior to the initial issue of facial pain - child had severe chicken pox and affected side had a number of noticable marks on that side of the face.

Number responding = 12 / percent responding = 8% of total respondents (143).

Page 65. IV. Triggers for Rombergs (page 2 of 2)

1. What possibly could have triggered Rombergs? (page 2):

Triggers	Responses (120)	% of Number responding
Symptoms start during puberty (both male and female)	29	38%
Symptoms accelerated during puberty (both male and female)	26	34%
Symptoms start during pregnancy	6	8%
Symptoms accelerated during pregnancy	8	10%
Changes in symptoms after child birth	16	21%
Symptoms start during menopause	1	1%
Symptoms accelerated during menopause	3	4%
Vitamin D deficiency	7	9%
My ideas for Other Triggers	24	32%

Number responding = 76 / percent responding = 53% of total respondents (143).
Note: This was a multiple choice question.

Page 66. IV. Triggers (page 2 of 2) cont.: Start during puberty
1. Please comment on the symptoms starting during puberty:

Responses (16)
The hair loss
Start age 11-12
When I first noticed my symptoms, I was a teen and had a darkening of skin near my mouth. According to Drs. who have seen my pictures of me as a child, they think that my mouth has always been crooked though??
Facial atrophy first became visible around the age of 11 and progressed for a few years, then stopping.
Diagnosed during puberty. At that stage I believe I already had atrophy to the face, back and chest. Began menstruating in late 2000.
Droopy eye lid and white patch of hair.
Dark circle under left eye at age 12, doctors thought I was anemic. Then after many doctor visits and at age 15 I was told that I had Romberg's.
Indentation down midforehead, also had severe acne, loss of bone in jaw.
First symptoms at age 10 - rapid progression at age 16 - 18 (not really during puberty but after start of puberty).
It's just that this is when it all started for her.
Also have ulcerative colitis, had the chicken pox at age 6.
Symptoms started age 19 years for me.
They started during puberty, but still think it has something to do with my car accident.
It became noticable at age 13. Only had 'smaller' right hand side of face and migraines. Facial pain, etc. started when I was 19.
Changes in face. Changes in color and more discoloration in affected area, more prominent indention in affected area, sever and debilitating migraine, vision problems, mental problems, learning problems. Low grade fever, extreme fatigue. Motion sickness. Intolerance to cold temeratures, very tough skin in all areas except Rombergs area where its thin. Severe constipation depending on mood.
My lip started to thin and my eye was droopy.

Number responding = 16 / percent responding = 11% of total respondents (143).

Page 67. IV. Triggers (page 2 of 2) cont.: Accelerated during puberty

1. Please comment on the symptoms accelerating during puberty:

Responses (18)
Changes in face, changes in color and more discoloration in affected area, more prominent indention in affected area, triggers, electrical impulses in brain, vision problems, mental problems, learning problems. Low grade fever and overwhelming feeling of being ill and tired. Extreme fatigue. Motion sickness. Intolerance to cold temperatures, very tough skin in all areas except Rombergs area where its thin. Changes in menses, constipation and abdominal pain depending on mood.
Noticed en coup de sabre during puberty, but had scalp lesion and hair loss at age 8.
If the tissue got hard in a few years ....it only needed months and there were significant changes because of the atrophy.
It seems the atrophy progressed rapidly as son approached puberty. I know that PRS is less common in males and therefore don't know if this experience w/puberty is common among other male patients. We originally thought my son "just had thin skin" on the right side of his chin. But as I mentioned before, it was as he approached puberty that the loss seemed to progress more rapidly and become more noticeable. From my reading of other's emails, it seems that many females affected by PRS discuss increase in symptoms during pregnancy and/or menopause. Seems to be a hormonal link.
I am not sure that the accelerating of symptoms during puberty has anything to do with puberty - my symptoms accelerated very much in my twenties too.
Progressed very slowly until 11-12 yrs., then accelerated from then on. We have other family members with other autoimmune diseases, not PRS.
As I got older the worse it became from what we (my family) can see.
Forceps birth??? Genetic???
Hormones
It started and accelerated when puberty started... just seems that when she did start to have an intermittent period, is when it progressed faster.
Continued atrophy of bone and soft tissue on forehead and jaw.
The medication lost effectivity and he got more and more seizures.
Seemed to increase.
According to doctors my mother took me to in my teens, my assymetry started in early childhood. During my teens, my mouth pulled further to one side, my cheek started to thin, my eye lashes turned white and fell out, I started to get migraines and jaw clamping, the crease in my chin started to get very noticable.
Seems that this was the time it was most noticeable because of my rapid growth spurts and the effected tissue not keeping up.
Could notice slight deterioration in my face in my teens. Mostly when I looked in a mirror.
Mouth was sore much of the time. I used wax on the wires.
Continued hair loss.

Number responding = 18 / percent responding = 13% of total respondents (143).

Page 68. IV. Triggers (page 2 of 2) cont.: Start during pregnancy

1. Please comment on the symptoms starting during pregnancy:

Responses (5)
Slight changes in my face began to appear during my second pregnancy and really accelerated after the birth.
It is only with hindsight that I realize I had PRS symptoms during pregnancy - third of four pregnancies was very difficult as I had mild paralysis on right side and was generally unwell.
Eye recession causing "droopy eyelid" after birth of 1st child. Aggravation during second pregnancy and after giving birth.
At first we thought I was retaining water and swollen on the other side, but after the birth of my child, we realized it was the other way around. No one was able to diagnosis me for 10 years and the disease progressed all through that time.
Vision problems, body feeling stiff all over, not able to walk or do anything, changes in face, changes in balance, thought and mood problems, left eye dilation after surgery, chronic constipation and fatigue, severe muscle and joint pain, low grade fever, headaches, overwhelming feeling of feeling ill, motion sickness, intolerance to cold temperatures, very tough hard skin in all areas except Rombergs area where it's thin, very small blood vessels, high blood pressure, borderline diabetic, total body pain and weakness, hypersensitivity to medications, blood work abnormal.

Number responding = 5 / percent responding = 3% of total respondents (143).

Page 69. IV. Triggers (page 2 of 2) cont.: Accelerated during pregnancy

1. Please comment on the symptoms accelerating during pregnancy:

Responses (6)
Noticed chin and jaw line deteriorating faster then before. Lost most of my top teeth for bone degeneration in my upper palette.
1st pregnancy: facial atrophy became more obvious, 2nd pregnancy: severe hip, back, knee and ankle problems so I was unable to walk.
Skin discolouration and more droopiness to eye lid.
Got worse with each pregnancy.
Noticed an increase in the atrophy during the time period of my 2 pregnancies in my early 20's.... have no idea if it was coincidental or causual.
Vision problems, body feeling stiff all over, not able to walk or do anything. Changes in face, changes in balance, thought and mood problems. Left eye dilation after surgery. Chronic constipation and fatigue. Severe muscle and joint pain, low grade fever, headaches. Overwhelming feeling of feeling ill. Motion sickness. Intolerance to cold temperatures, very tough hard skin in all areas except Rombergs area where it's thin. Very small blood vessels, high blood pressure. Borderline diabetic. Total body pain and weakness. Hypersensitivity to medications. Blood work abnormal.

Number responding = 6 / percent responding = 4% of total respondents (143).

Page 70. IV. Triggers (page 2 of 2) cont.: Changes in symptoms after child birth

1. Please comment on the changes in symptoms after child birth:

Responses (12)
Vision problems, Body feeling stiff all over, not able to walk or do anything. Changes in face, Changes in balance, thought and mood problems. Left eye dilation after surgery. Chronic constipation and fatigue. severe muscle and joint pain, low grade fever. headaches. Overwhelming feeling of feeling ill. Motion sickness. Intolerance to cold temperatures, very tough hard skin in all areas except Rombergs area where it's thin. Very small blood vessels, high blood pressure, borderline diabetic. Total body pain and weakness. Problems with kidney and kidney stones. Hypersensitivity to medications, Blood work abnormal.
After I had my son, when he was about 4 years old, that is when I noticed the problem.
The right side of my face started "melting away". It was at that time that I first sought treatment for the problem.
Hard to say if it was during pregnancy or after child birth....but for sure things got worse each time.
I lost my teeth prior to my pregnancy but it seems that most of the progression happened after the birth of my son.
Had first baby at age 28. Line appeared on nose, first noticed after baby born.
Has continued to progress since that time.
Noticed that symptoms increased after child birth.
I gave birth 7 months ago. I feel as though the scleroderma and PR has reactivated. I have definite spots of discoloration on my arms and torso that I did not have prior to pregnancy. I also have pain in my face. I believe it is the combination of hormones, stress and lack of sleep causing this progression.
I did have surgery right after my daughter was born- fat injections, but it could have been just the timing of when I could do it. I guess I cannot actually say for certain right now that I can remember a definite change from the times when I did have kids. sorry!
Tissue loss was more pronounced, before only bone structure had been changing now, then tissue was degenerating.
Discoloration, indentation, increased facial atrophy.

Number responding = 12 / percent responding = 8% of total respondents (143).

Page 71. IV. Triggers (page 2 of 2) cont.: Start during menopause

1. Please comment on the symptoms starting during menopause:

Responses (1)
Hindsight again - knew very little about PRS until joining the group, but started to get shingles during menopause.

Number responding = 1 / percent responding = 0.7% of total respondents (143).

Page 72. IV. Triggers (page 2 of 2) cont.: Accelerated during menopause

1. Please comment on the symptoms accelerating during menopause:

Responses (3)
I had a clear second round of Rombergs in my late 40's as I was entering parimenopause. It included loss of teeth, eyelid twitching, discoloration of forehead skin. Romberg started lower on my face, around my jaw and cheek, and traveled upward. The en coup de sabre crossed my eye in my late 40's as well.
First I noticed discolorations in my forehead. For several months I've been experiencing a burning sensation in the left forehead area and nose. I'm not sure, but I think I'm noticing further tissue loss on the left side of my nose. Too early to know for sure, but I am monitoring.
Just started up more rapidly with menopause.

Number responding = 3 / percent responding = 2% of total respondents (143).

Page 73. IV. Triggers (page 2 of 2) cont.: Vitamin D deficiency
1. Please comment on a Vitamin D deficiency:

Responses (6)
Very possible
I wonder if mine is caused by SPF (sun protection cream) use because I use it to keep dark lines from getting darker. Have been on supplements for awhile now.
I now have vitamin D deficiency. I'm not sure if it was related to the IV steroid treatment I had or if I have always had it but didn't know it. I'm taking supplements but the progression still continues.
My Dr. first diagnosed me with very low vitamin D (diagnosed at 7) about a year and a half ago. That is the first time any Dr. had ever checked for vitamin D deficiency. I was prescribed 50,000 U once a week for quite a while (until my vitamin D was normal), then prescribed 50,000 U once a month.
I take vitamin D now and I think it helps. I have the girls take it as well.
I recently discovered I am severly vitamin D deficient.

Number responding = 6 / percent responding = 4% of total respondents (143).

Page 74. IV. Triggers (page 2 of 2) cont.: Other Triggers

1. Please comment on your "Other" ideas for Romberg Triggers:

Responses (22)
Trauma from car accident accelerated or re-started symptoms.
Trauma caused by dental work.
Because pregnancy was not confirmed until 5th month, I continued to take birth control pills. Forceps delivery.
I have no idea, nor do my parents, for a cause or trigger as to why I was affected by Rombergs.
As stated in other areas of this survey, I believe that the head trauma experienced at age 5 might have triggered PRS. I also, believe the trauma of having my wisdom teeth pulled at 24 reactivated it.
Head injuries, plus strong family history of autoimmunity. Pregnancies were accelerants.
Had a very difficult birth. Doctor used the suction to extract him... 31 times! Unheard of! He had migraines at birth and was not breathing at first. Emergency situation.
I'm not, I do believe stress makes it worse.
I had taken a series of vaccination against Candida albicans, I was under strong stress at work, I had Herpes Zoster. And I had a history of Thyroid autoimmune disease.
Could be taking out the molar or the accident with my forehead affected.
About two weeks before we noticed the first sign of Rombergs, I had the mumps.
Still suspect that the head injury approximately one year prior to the start of my PR diagnoisis may have been the trigger.
Trauma to the initial side of forehead was beginning of disease process.
Fell on cement stairs.
Injury and virus caused my immune system to become confused. So instead of attacking the virus, it attacked the injured area instead, triggering Rombergs.
Did other people with Rombergs eat a lot of processed/microwaved foods? Patrick loves Michelinas Wheels and Cheese and I regret that I let him eat many, many boxes that were microwaved in the plastic lined box. Has anyone else reported picky eaters who consumed too much of one food? Or, any other food/diet issues?
Forceps delivery. As this head trauma could have affected my eyes as well as face. There was a mark left from the forceps when I was born over the affected side/eye.
Injuries from difficult birth. Possible exposure to ticks as a toddler.(?)
Had a tooth crowned. That very day I noticed the droop in her lip.
As a teenager, I was under quite a bit of emotional stress at the time of tissue loss. It seems to me that each decade of my life has seen a different complication. The initial trauma at ten, major tissue loss at 15, orbital tissue loss at 27, vision complications during 30's, seizures during 40's, I'm 55, I have noticed lack of coordination, thought it was age related.
Sexually abused as a kid, disease started one to two years after. I believe there is a genetic predisposition which was triggered by stress. Mind and body are related.
I had a bad pregnancy and labor with her. She had a rough delivery. Cord around her neck twice she was being a yoyo. She was hitting my pelvis. Bad. We almost died.

Number responding = 22 / percent responding = 15% of total respondents (143).

Page 75. V. Treatment for Rombergs

1. Please select any treatments that have been used:

Treatments	Responses (128)	% of Number responding
Medications	43	44%
Other types of treatment	25	26%
Surgery	60	62%

Number responding = 97 / percent responding = 68% of total respondents (143).
Note: This was a multiple choice question.

Page 76. V. Treatment for Rombergs cont.: Medications Used (page 1 of 13)

1. Select any medications used:

Medication	Responses (67)	% of Number responding
Aspirin	6	18%
Diprosone	1	3%
Doryx	0	0%
Dovonex	3	9%
Doxycycline	1	3%
Gabapentin	2	6%
Intravenous Gamma Globulin (IVIG)	2	6%
Methotrexate	18	53%
Methylprednisolone	3	9%
Monocycline	0	0%
Oral Calcitriol	1	3%
Penicillamine	2	6%
Plaquenil	3	9%
Prednisolone	6	18%
Prednisone	10	29%
Quinacrine	1	3%
Topical antibiotics (Bacitricin - Neosporin)	4	12%
vitamin B-12	4	12%

Number responding = 34 / percent responding = 24% of total respondents (143).
Note: This was a multiple choice question.

Page 76. V. Treatment for Rombergs cont.: Medications Used (page 1 of 13)

2. For "Other" medications, please enter:

Medication	Responses (56)	% of Number responding
2.9 gram Rocephin (antibiotics) daily, during 30 days	1	5%
Adderall	1	5%
Albuterol	2	9%
Anti seizure meds	5	23%
Ceftriaxone	1	5%
Clobetasol Propionate cream	1	5%
Codeine	1	5%
Colchicina	1	5%
Colchicine (colchis)	1	5%
Compazine	1	5%
Corticoizi	1	5%
Darvocet	1	5%
eye drops	1	5%
Flexeril	1	5%
folic acid	3	14%
I do not know type - series of steroid injections to face during first year - no effect	1	5%
iron tablets	1	5%
Keppra	2	9%
Lexapro, Zoloft, Effexor for depression now Seroquil possibly	1	5%
Liquid silicone injections	1	5%
Midrin as needed	1	5%
Migratin?	1	5%
Moldamin 1200000 UI	1	5%
MS Contin for pain	1	5%
Paracetamol	1	5%
Pax (Valium)2	1	5%
Penicillin	1	5%

Continued on the next page.

Medication	Responses (56)	% of Number responding
Petadolex (my daughter takes for her migraine headaches)	1	🟦 5%
Piascledine 300	1	🟥 5%
Prednisone one time prescription for headache	1	🟦 5%
Prescription Aleve as needed	1	🟥 5%
Prilosec	2	🟦 9%
Riddalin	1	🟥 5%
Salazaphrine	1	🟦 5%
STEROIDS, BUT THE NAME IS UNKNOWN	1	🟥 5%
Tamal	1	🟦 5%
various pain relievers	1	🟥 5%
Vicodin	1	🟦 5%
vitamin C intravenous therapy	1	🟥 5%
vitamin D	1	🟦 5%
vitamin D & calcium	1	🟥 5%
Vitix	1	🟦 5%
Voltarin	1	🟥 5%
was on Prednisone for the colitus before the onset of prs	1	🟦 5%
Xanax for the neuropathy	1	🟥 5%
Zantac	2	🟦 9%

Number responding = 22 / percent responding = 15% of total respondents (143).
Note: This was a multiple choice question.

Note: For the rest of this Medication section, only the first few words of the "Other Medications" will be shown.

Page 77. V. Treatment for Rombergs cont.: Medications Used (page 2 of 13)

1. Is the affected person taking this medication orally or by injection?

Medication	Responses (102)	Orally	Injection	(IV) Intravenous
Aspirin	4	100% (4)		
Diprosone	1	100% (1)		
Doryx	0			
Dovonex	0			
Doxycycline	1	100% (1)		
Gabapentin	1	100% (1)		
Intravenous Gamma Globulin (IVIG)	1			100% (1)
Methotrexate	19	68% (13)	31% (6)	
Methylprednisolone	2			100% (2)
Monocycline	0			
Oral Calcitriol	1	100% (1)		
Penicillamine	1			100% (1)
Plaquenil	3	100% (3)		
Prednisolone	7	71% (5)		28% (2)
Prednisone	8	100% (8)		
Quinacrine	0			
Topical antibiotics	2	50% (1)	50% (1)	
vitamin B-12	4	100% (1)		
List of "Other Medications" Follows				
2.9 gram Rocephin…	1			100% (1)
Adderall	1	100% (1)		
Albuterol	1	100% (1)		
Albuterol	1	100% (1)		
anti seizure meds	1	100% (1)		
anti seizure meds	1	100% (1)		
anti seizure meds	1	100% (1)		
anti seizure meds	1	100% (1)		
anti seizure meds	1	100% (1)		
Ceftriaxone	1			100% (1)
Clobetasol Propionate cream	1			
Codeine	1	100% (1)		
Colchicina	1	100% (1)		
Colchicine (colchis)	1	100% (1)		
Compazine	1	100% (1)		
Corticoizi	1	100% (1)		

Continued on the next page.

Medication	Responses (102)	Orally	Injection	(IV) Intravenous
Darvocet	1	100% (1)		
eye drops	1	100% (1)		
Flexeril	1	100% (1)		
folic acid	1	100% (1)		
folic acid	1	100% (1)		
folic acid	1	100% (1)		
I do not know type…	1			
iron tablets	1			
Keppra	1	100% (1)		
Keppra	1	100% (1)		
Lexapro, Zoloft, Effexor…	1			
Liquid silicone injections	1			
Midrin as needed	1			
Migratin?	1			
Moldamin 1200000 UI	1		100% (1)	
MS Contin for pain	1	100% (1)		
Paracetamol	1	100% (1)		
Pax (Valium)	1	100% (1)		
Penicillin	1			100% (1)
Petadolex (my daughter…	1	100% (1)		
Piascledine 300	1	100% (1)		
Prednisone one time…	1			
Prescription Aleve as needed	1			
Prilosec	1	100% (1)		
Prilosec	1	100% (1)		
Riddalin	1	100% (1)		
Salazaphrine	1	100% (1)		
STEROIDS, BUT…	1			
Tamal	1	100% (1)		
various pain relievers	1	100% (1)		
Vicodin	1	100% (1)		
vitamin C intravenous therapy	1			100% (1)
vitamin D	1	100% (1)		
vitamin D & calcium	1	100% (1)		
Vitix	1	100% (1)		
Voltarin	1		100% (1)	
was on Prednisone…	1	100% (1)		

Continued on the next page.

Xanax for the neuropathy	1	100% (1)		
Zantac	1	100% (1)		
Zantac	1	100% (1)		
Medication	**Responses (102)**	**Orally**	**Injection**	**(IV) Intravenous**

Number responding = 37 / percent responding = 26% of total respondents (143).
Note: This was a multiple choice question.

Page 78. V. Treatment for Rombergs cont.: Medications Used (page 3 of 13)

1. At what age was this medication started?

Medication	Responses (120)	Age When This Medication Was Started
Aspirin	6	In my late 50's, 6, child, 40's, not, 30
Diprosone	2	About 10 or 11, not
Doryx	1	not
Dovonex	3	44-only used it for a week because rash all over chin broke out 26 not
Doxycycline	2	originally for 3 weeks after lyme, then put on again for 3 months 3 years later not
Gabapentin	2	42 - on it for 4 months but this is when started twitching, so went off it. Continued twitching for a year plus not
Intravenous Gamma Globulin (IVIG)	2	14, not
Methotrexate	19	7, 6, 7, 7.5 yrs to 9 yrs., 5, 4, 9, 43, 13, 8, 26, age 18, age 7, 13, 43, 9, 14, not, 31
Methylprednisolone	3	5, 45, not
Monocycline	1	not
Oral Calcitriol	2	13, not
Penicillamine	2	29, not
Plaquenil	4	20, age 17, age 7, not
Prednisolone	5	7, 13, 8, 9, not
Prednisone	11	7, 41, 3, 26, age 5, 13, 7 20's tried the oral and the eye drops for a short period of time. No longer taking it. around 16, not, 32
Quinacrine	2	18, not
Topical antibiotics	3	It could be imagination, but sometimes area looks better after having Neosporin on it overnight. 13, not
List of "Other Medications" Follows		
2.9 gram Rocephin...	1	7
Adderall	1	30
Albuterol	1	7
Albuterol	1	7
anti seizure meds	1	
anti seizure meds	1	
anti seizure meds	1	

Continued on the next page.

Medication	Responses (120)	Age When This Medication Was Started
anti seizure meds	1	
anti seizure meds	1	
Ceftriaxone	1	
Clobetasol Propionate	1	13
Codeine	1	29
Colchicina	1	16
Colchicine (colchis)	1	43
Compazine	1	30
Corticoizi	1	13
Darvocet	1	20's
eye drops	1	30
Flexeril	1	29
folic acid	1	13
Folic acid	1	7
folic acid	1	7
I do not know type…	1	
iron tablets	1	7
Keppra	1	7
Keppra	1	7
Lexapro, Zoloft, Effexor…	1	
Liquid silicone injections	1	
Midrin as needed	1	
Migratin?	1	
Moldamin 1200000 UI	1	13
MS Contin for pain	1	38
Paracetamol	1	22
Pax (Valium)	1	22
Penicillin	1	41
Petadolex (my daughter…	1	17 (my daughter)
Piascledine 300	1	16
Prednisone one time…	1	
Prescription Aleve as needed	1	
Prilosec	1	7
Prilosec	1	7
Riddalin	1	25
Salazaphrine	1	7

Continued on the next page.

Medication	Responses (120)	Age When This Medication Was Started
STEROIDS, BUT…	1	AGE 7
Tamal	1	22
various pain relievers	1	puberty
Vicodin	1	40's
vitamin C intravenous therapy	1	27
vitamin D	1	50
vitamin D & calcium	1	13
Vitix	1	18
Voltarin	1	22
was on Prednisone…	1	7
Xanax for the neuropathy	1	38
Zantac	1	7
Zantac	1	7

Number responding = 38 / percent responding = 27% of total respondents (143).
Note: This was a multiple choice question.

Page 79. V. Treatment for Rombergs cont.: Medications Used (page 4 of 13)

1. If the medication has been stopped, what was the age of the person?

Medication	Responses (65)	Age When This Medication Was Stopped
Aspirin	3	20, not, still taking
Diprosone	2	13, not
Doryx	1	not
Dovonex	2	not 44-only used it for a week because rash all over chin broke out
Doxycycline	2	not, 43
Gabapentin	2	not, 42
Intravenous Gamma Globulin (IVIG)	2	14, not
Methotrexate	7	not 32, 15, 6 9 yrs. - stopped due to upcoming surgery 7, 43
Methylprednisolone	3	45, 5.5, not
Monocycline	1	not
Oral Calcitriol	2	14, not
Penicillamine	2	not, 29
Plaquenil	3	not, age 18, 21
Prednisolone	4	8, 9, 13, not
Prednisone	6	34, 41, 20's, 16, around 16, not
Quinacrine	2	not, 21
Topical antibiotics	2	not, 13
vitamin B-12	2	not, 41
List of "Other Medications" Follows		
2.9 gram Rocephin…	1	7
Adderall		
Albuterol		
Albuterol		
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
Ceftriaxone		
Clobetasol Propionate		
Codeine		
Colchicina	1	19

Continued on the next page.

Medication	Responses (65)	Age When This Medication Was Stopped
Colchicine (colchis)	1	43
Compazine		
Corticoizi		
Darvocet		
eye drops		
Flexeril		
folic acid	1	13
Folic acid	1	13
folic acid		
I do not know type…		
iron tablets	1	16
Keppra		
Keppra		
Lexapro, Zoloft, Effexor…		
Liquid silicone injections		
Midrin as needed		
Migratin?		
Moldamin 1200000 UI	1	18
MS Contin for pain		
Paracetamol		
Pax (Valium)		
Penicillin	1	41
Petadolex (my daughter…	1	still taking
Piascledine 300	1	15
Prednisone one time…		
Prescription Aleve as needed		
Prilosec		
Prilosec		
Riddalin		
Salazaphrine	1	16
STEROIDS, BUT…	1	12
Tamal		
various pain relievers	1	puberty
Vicodin		
vitamin C intravenous therapy	1	27
vitamin D	1	still taking
vitamin D & calcium		

Continued on the next page.

Vitix	1	16
Voltarin		
was on Prednisone…	1	16
Xanax for the neuropathy		
Zantac		
Zantac		
Medication	**Responses (65)**	**Age When This Medication Was Stopped**

Number responding = 38 / percent responding = 27% of total respondents (143).
Note: This was a multiple choice question.

Page 80. V. Treatment for Rombergs cont.: Medications Used (page 5 of 13)

1. Did this medication stop the atrophy, reverse the atrophy or have no change for the atrophy:

Medication	Responses (105)	Stop	Reverse	No change
Aspirin	6			100% (6)
Diprosone	2	50% (1)		50% (1)
Doryx	1			100% (1)
Dovonex	3			100% (3)
Doxycycline	1			100% (1)
Gabapentin	1			100% (1)
Intravenous Gamma Globulin (IVIG)	2			100% (2)
Methotrexate	17	33% (6)		61% (11)
Methylprednisolone	3	33% (1)		66% (2)
Monocycline	1			100% (1)
Oral Calcitriol	2	100% (1)		100% (1)
Penicillamine	1			100% (1)
Plaquenil	4			100% (4)
Prednisolone	6	33% (2)		66% (4)
Prednisone	9	11% (1)		88% (8)
Quinacrine	2			100% (2)
Topical antibiotics	2			100% (2)
vitamin B-12	4			100% (4)
List of "Other Medications" Follows				
2.9 gram Rocephin...	1	100% (1)		
Adderall	1			100% (1)
Albuterol	1			100% (1)
Albuterol	1			100% (1)
anti seizure meds				
anti seizure meds				
anti seizure meds				
anti seizure meds				
anti seizure meds				
Ceftriaxone	1			100% (1)
Clobetasol Propionate cream	1			
Codeine				
Colchicina	1			100% (1)
Colchicine (colchis)	1	100% (1)		
Compazine	1			100% (1)
Corticoizi	1			100% (1)

Continued on the next page.

Medication	Responses (105)	Stop	Reverse	No change
Darvocet	1			100% (1)
eye drops				
Flexeril				
folic acid	1			100% (1)
folic acid	1			100% (1)
folic acid	1			100% (1)
I do not know type…				
iron tablets	1			100% (1)
Keppra	1			100% (1)
Keppra	1			100% (1)
Lexapro, Zoloft, Effexor…				
Liquid silicone injections				
Midrin as needed				
Migratin?				
Moldamin 1200000 UI	1			100% (1)
MS Contin for pain	1			100% (1)
Paracetamol	1			100% (1)
Pax (Valium)	1			100% (1)
Penicillin	1			100% (1)
Petadolex (my daughter…				
Piascledine 300	1			100% (1)
Prednisone one time…				
Prescription Aleve as needed				
Prilosec	1			100% (1)
Prilosec	1			100% (1)
Riddalin				
Salazaphrine	1			100% (1)
STEROIDS, BUT…	1			100% (1)
Tamal	1			100% (1)
various pain relievers	1			100% (1)
Vicodin	1			100% (1)
vitamin C intravenous therapy	1		100% (1)	
vitamin D				
vitamin D & calcium	1			100% (1)
Vitix	1			100% (1)
Voltarin	1			100% (1)
was on Prednisone…	1			100% (1)

Continued on the next page.

Xanax for the neuropathy	1			100% (1)
Zantac	1			100% (1)
Zantac	1			100% (1)
Medication	**Responses (105)**	**Stop**	**Reverse**	**No change**

Number responding = 34 / percent responding = 24% of total respondents (143).
Note: This was a multiple choice question.

Page 81. V. Treatment for Rombergs cont.: Medications Used (page 6 of 13)

1. If there were any improvements, how long before you noticed them?

Medication	Responses (32)	Any Improvements, How Long Before You Noticed
Aspirin	2	not, used to prevent vascular constriction
Diprosone		
Doryx		
Dovonex		
Doxycycline		
Gabapentin		
Intravenous Gamma Globulin (IVIG)		
Methotrexate	6	none 3 months slowed it down 5 months the disease stopped after a while, can't say it was because of the drug 2-3 months
Methylprednisolone	1	2-3 months
Monocycline		
Oral Calcitriol	1	2 - 3 months seemed to slow it down
Penicillamine		
Plaquenil		
Prednisolone	2	none, approx. 2 weeks into treatment
Prednisone	3	Immediate note this taken for ulcerative colitus prior to PRS so it made no difference to PRS 3 months
Quinacrine		
Topical antibiotics		
vitamin B-12	1	none
List of "Other Medications" Follows		
2.9 gram Rocephin…		
Adderall		
Albuterol		
Albuterol	1	none
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		

Continued on the next page.

Medication	Responses (32)	Any Improvements, How Long Before You Noticed
anti seizure meds		
Ceftriaxone		
Clobetasol Propionate		
Codeine		
Colchicina	1	19
Colchicine (colchis)	1	the disease stopped after a while, can't say it was because of the drug
Compazine		
Corticoizi		
Darvocet		
eye drops		
Flexeril		
folic acid	1	13
Folic acid	1	13
folic acid	1	none
I do not know type…		
iron tablets	1	note this taken for ulcerative colitus prior to PRS so it made no difference to PRS
Keppra		
Keppra	1	none
Lexapro, Zoloft, Effexor…		
Liquid silicone injections		
Midrin as needed		
Migratin?		
Moldamin 1200000 UI		
MS Contin for pain		
Paracetamol		
Pax (Valium)		
Penicillin		
Petadolex (my daughter…	1	within a week
Piascledine 300		
Prednisone one time…		
Prescription Aleve as needed		
Prilosec		
Prilosec	1	none
Riddalin		

Continued on the next page.

Medication	Responses (32)	Any Improvements, How Long Before You Noticed
Salazaphrine	1	note this taken for ulcerative colitus prior to PRS so it made no difference to PRS
STEROIDS, BUT…		
Tamal		
various pain relievers	1	immediate pain relief
Vicodin		
vitamin C intravenous therapy	1	2 months
vitamin D	1	I think that vitamin D has slowed down the atrophic process
vitamin D & calcium		
Vitix		
Voltarin		
was on Prednisone…	1	note this taken for ulcerative colitus prior to PRS so it made no difference to PRS
Xanax for the neuropathy		
Zantac		
Zantac	1	none

Number responding = 12 / percent responding = 8% of total respondents (143).
Note: This was a multiple choice question.

1. If there are or were any side affects, please list them:

Medication	Responses (38)	Any Side Affects, Please List Them
Aspirin	1	not
Diprosone	1	weekly blood counts as my lips were always blue
Doryx		
Dovonex		
Doxycycline		
Gabapentin	1	body wide twitching/fasciculations started
Intravenous Gamma Globulin (IVIG)		
Methotrexate	10	nausea Candida (it came back after a series of vaccination - my immunity was very low) vomiting and nausea cranky for 24 hours after weekly injection nausea absolutely no side affects noticed sores on lips - counteracted with folic acid extreme fatigue nausea and fatigue none
Methylprednisolone	2	nausea nausea, fatigue, fluid retention, dizziness, racing heart
Monocycline		
Oral Calcitriol		
Penicillamine		
Plaquenil		
Prednisolone	3	Mood change, hunger, exhaustion weight gain tiredness, huge appetite, tearful
Prednisone	4	minor moon face gain 30 kg hyperactivity
Quinacrine		
Topical antibiotics		
vitamin B-12	1	none
List of "Other Medications" Follows		
2.9 gram Rocephin...		
Adderall		
Albuterol		

Continued on the next page.

Medication	Responses (38)	Any Side Affects, Please List Them
Albuterol	1	none
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
Ceftriaxone		
Clobetasol Propionate	1	Quit using after about 1 1/2 months. Scalp was coming up in chunks.
Codeine		
Colchicina	1	skin rash... acne
Colchicine (colchis)		
Compazine		
Corticoizi		
Darvocet		
eye drops		
Flexeril		
folic acid		
Folic acid		
folic acid	1	none
I do not know type…		
iron tablets		
Keppra		
Keppra	1	none
Lexapro, Zoloft, Effexor…		
Liquid silicone injections		
Midrin as needed		
Migratin?		
Moldamin 1200000 UI	1	stomach problems
MS Contin for pain	1	constipation, dizziness
Paracetamol		
Pax (Valium)		
Penicillin	1	Sludge in gall bladder
Petadolex (my daughter…		
Piascledine 300	1	muscular pain and problems with walking
Prednisone one time…		
Prescription Aleve as needed		

Continued on the next page.

Medication	Responses (38)	Any Side Affects, Please List Them
Prilosec		
Prilosec	1	none
Riddalin		
Salazaphrine		
STEROIDS, BUT…	1	SKIN ERUPTIONS, BECAME VERY MUSCULAR
Tamal		
various pain relievers	1	constipation
Vicodin		
vitamin C intravenous therapy	1	interfered with methotrexate
vitamin D		
vitamin D & calcium		
Vitix	1	stomach problems
Voltarin		
was on Prednisone…		
Xanax for the neuropathy	1	sleepiness
Zantac		
Zantac	1	none

Number responding = 22 / percent responding = 15% of total respondents (143).
Note: This was a multiple choice question.

Page 83. V. Treatment for Rombergs cont.: Medications Used (page 8 of 13)

1. If there are or were side affects, how debilitating are or were they?

Medication	Responses (28)	How Debilitating Are or Were They
Aspirin	1	not
Diprosone		
Doryx		
Dovonex		
Doxycycline		
Gabapentin		
Intravenous Gamma Globulin (IVIG)		
Methotrexate	4	he felt sick for 1 - 2 days none It took 6 - 9 months to adjust to this medication. Still fatigued though. not sure
Methylprednisolone	2	He was sick for about a week after each dose. It was horrible. I did 3 days in 3 separate months. By the 3rd day, I felt like I was going to collapse.
Monocycline		
Oral Calcitriol		
Penicillamine		
Plaquenil		
Prednisolone	2	none Missed school, but wore off in a few days
Prednisone	4	minor can't really remember too much weight gain, mood swings - we are ready for her to be off of it, & she is coming off it slowly very
Quinacrine		
Topical antibiotics		
vitamin B-12	1	none
List of "Other Medications" Follows		
2.9 gram Rocephin…		
Adderall		
Albuterol		
Albuterol	1	none
anti seizure meds		
anti seizure meds		
anti seizure meds		

Continued on the next page.

Medication	Responses (28)	How Debilitating Are or Were They
anti seizure meds		
anti seizure meds		
Ceftriaxone		
Clobetasol Propionate	1	not debilitating just irritating and gross
Codeine		
Colchicina		
Colchicine (colchis)		
Compazine		
Corticoizi		
Darvocet		
eye drops		
Flexeril		
folic acid	1	none
Folic acid		
folic acid	1	none
I do not know type…		
iron tablets		
Keppra		
Keppra	1	none
Lexapro, Zoloft, Effexor…		
Liquid silicone injections		
Midrin as needed		
Migratin?		
Moldamin 1200000 UI	1	very
MS Contin for pain	1	have learned to live with them
Paracetamol		
Pax (Valium)		
Penicillin		
Petadolex (my daughter…		
Piascledine 300	1	very
Prednisone one time…		
Prescription Aleve as needed		
Prilosec		
Prilosec	1	none
Riddalin		
Salazaphrine		

Continued on the next page.

Medication	Responses (28)	How Debilitating Are or Were They
STEROIDS, BUT…	1	THE STEROIDS CHANGED MY PERSONALITY
Tamal		
various pain relievers	1	not very debilitating
Vicodin		
vitamin C intravenous therapy		
vitamin D		
vitamin D & calcium	1	none
Vitix		
Voltarin		
was on Prednisone…		
Xanax for the neuropathy	1	have learned to live with them
Zantac		
Zantac	1	none

Number responding = 12 / percent responding = 8% of total respondents (143).
Note: This was a multiple choice question.

Page 84. V. Treatment for Rombergs cont.: Medications Used (page 9 of 13)

1. If any testing was advised while on this medication, please list these tests:

Medication	Responses (21)	Any Testing Advised, Please List
Aspirin	1	not
Diprosone	1	Regular blood count
Doryx		
Dovonex		
Doxycycline		
Gabapentin		
Intravenous Gamma Globulin (IVIG)		
Methotrexate	12	blood testings blood tests to make sure liver function, etc., is ok blood work every 3 months testing toxicity to liver functions blood tests Blood count and liver enzyme test done monthly liver function monthly blood work blood test bloods every 5 weeks, hospital check up every 12 weeks Regular blood test LFT (Liver Function Tests) blood chem monthly blood and urine tests
Methylprednisolone	2	blood test every day while having the IV, LFT (Liver Function Tests) blood work
Monocycline		
Oral Calcitriol	1	calcium levels monthly
Penicillamine		
Plaquenil	1	monthly blood and urine tests
Prednisolone	1	urine and blood pressure weekly
Prednisone	2	monthly blood and urine tests blood tests
Quinacrine		
Topical antibiotics		
vitamin B-12		
List of "Other Medications" Follows		
2.9 gram Rocephin…		
Adderall		
Albuterol		
Albuterol		

Continued on the next page.

Medication	Responses (21)	Any Testing Advised, Please List
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
Ceftriaxone		
Clobetasol Propionate		
Codeine		
Colchicina		
Colchicine (colchis)		
Compazine		
Corticoizi		
Darvocet		
eye drops		
Flexeril		
folic acid		
Folic acid		
folic acid		
I do not know type…		
iron tablets		
Keppra		
Keppra		
Lexapro, Zoloft, Effexor…		
Liquid silicone injections		
Midrin as needed		
Migratin?		
Moldamin 1200000 UI		
MS Contin for pain		
Paracetamol		
Pax (Valium)		
Penicillin		
Petadolex (my daughter…		
Piascledine 300		
Prednisone one time…		
Prescription Aleve as needed		
Prilosec		

Continued on the next page.

Medication	Responses (21)	Any Testing Advised, Please List
Prilosec		
Riddalin		
Salazaphrine		
STEROIDS, BUT…		
Tamal		
various pain relievers		
Vicodin		
vitamin C intravenous therapy		
vitamin D		
vitamin D & calcium		
Vitix		
Voltarin		
was on Prednisone…		
Xanax for the neuropathy		
Zantac		
Zantac		

Number responding = 14 / percent responding = 10% of total respondents (143).
Note: This was a multiple choice question.

Page 85. V. Treatment for Rombergs cont.: Medications Used (page 10 of 13)

1. If the medication was stopped, please explain why it was stopped:

Medication	Responses (38)	Please Explain Why It Was Stopped
Aspirin	1	not
Diprosone	1	Had been taking it for awhile so it was decided to stop to see what the outcome would be.
Doryx		
Dovonex	1	rash
Doxycycline	1	Doctor said couldn't take it anymore as long term use can trigger lupus and I already had positive ANA and he thought Lyme was treated.
Gabapentin	1	twitching was unnerving to me
Intravenous Gamma Globulin (IVIG)	1	no effects
Methotrexate	4	Thought it had done the job and it should be stopped as soon as posible. did not see enough signs PRS stopped the idea was to take it just for a few months We are going to have fat injections done in April 2009 so we were advised to stop.
Methylprednisolone	2	The plan was to hit the body hard with the steroids. The doctor prescribed a 3 IV steroid treatments each month for 3 months in a row. set amount over 6 months
Monocycline		
Oral Calcitriol	1	stopped seeing changes
Penicillamine		
Plaquenil	2	severe stomach upset was not working
Prednisolone	2	treatment was 3 infustions daily for 3 days in a row lasting 4 months end of planned treatment
Prednisone	4	generally wasn't comfortable with steroids This was for treating ulcerative colitus - went on lactose free diet which helped. no improvement cortisone pulse - therapy
Quinacrine	1	changed country and no effect
Topical antibiotics	1	no improvement
vitamin B-12	1	no change
List of "Other Medications" Follows		
2.9 gram Rocephin...	1	The medication was given because of the Lyme infection, but after this medication was taken headaches became milder and tissue disappearance became much less/stopped. But it is of course also possible this improvement is not related to the medication but......
Adderall		
Albuterol		
Continued on the next page.		

Medication	Responses (38)	Please Explain Why It Was Stopped
Albuterol	1	also takes Naproxen
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
anti seizure meds		
Ceftriaxone		
Clobetasol Propionate	1	because of what it was doing to her skin
Codeine		
Colchicina	1	no improvement
Colchicine (colchis)	1	the idea was to take it just for a few months
Compazine		
Corticoizi	1	no improvement
Darvocet		
eye drops		
Flexeril		
folic acid		
Folic acid		
folic acid		
I do not know type…		
iron tablets	1	His was for treating Ulcerative Colitus - went on lactose free diet which helped.
Keppra		
Keppra		
Lexapro, Zoloft, Effexor…		
Liquid silicone injections		
Midrin as needed		
Migratin?		
Moldamin 1200000 UI	1	no improvement
MS Contin for pain		
Paracetamol		
Pax (Valium)		
Penicillin	1	pulse-therapy, planned duration for 3 weeks, 20 Mio. units per day
Petadolex (my daughter…		
Piascledine 300		

Continued on the next page.

Medication	Responses (38)	Please Explain Why It Was Stopped
Prednisone one time…		
Prescription Aleve as needed		
Prilosec		
Prilosec		
Riddalin		
Salazaphrine	1	His was for treating Ulcerative Colitus - went on lactose free diet which helped.
STEROIDS, BUT…	1	BECAUSE IT HAD NOT HELPED THE ROMBERGS AND WAS CREATING OTHER ISSUES.
Tamal		
various pain relievers		
Vicodin		
vitamin C intravenous therapy	1	interfered with methotrexate
vitamin D		
vitamin D & calcium		
Vitix	1	no improvement
Voltarin		
was on Prednisone…	1	His was for treating Ulcerative Colitus - went on lactose free diet which helped.
Xanax for the neuropathy		
Zantac		
Zantac		

Number responding = 22 / percent responding = 15% of total respondents (143).
Note: This was a multiple choice question.

Page 86. V. Treatment for Rombergs cont.: Medications Used (page 11 of 13)

1. If the medication was stopped, did any of the symptoms return after a period of time?

Medication	Responses (52)	Yes	No	Undecided
Aspirin	2		50% (1)	50% (1)
Diprosone	2		50% (1)	50% (1)
Doryx	1			100% (1)
Dovonex	1			100% (1)
Doxycycline	1			100% (1)
Gabapentin	2	50% (1)		50% (1)
Intravenous Gamma Globulin (IVIG)	1			100% (1)
Methotrexate	5		40% (2)	60% (3)
Methylprednisolone	3	33% (1)		66% (2)
Monocycline	1			100% (1)
Oral Calcitriol	2			100% (2)
Penicillamine	2	100% (1)		100% (1)
Plaquenil	3	66% (2)		33% (1)
Prednisolone	2		50% (1)	50% (1)
Prednisone	6	50% (3)	16% (1)	33% (2)
Quinacrine	1			100% (2)
Topical antibiotics	1	100% (1)		
vitamin B-12	2	50% (1)		50% (1)
List of "Other Medications" Follows				
2.9 gram Rocephin...				
Adderall				
Albuterol				
Albuterol				
anti seizure meds				
anti seizure meds				
anti seizure meds				
anti seizure meds				
anti seizure meds				
Ceftriaxone				
Clobetasol Propionate cream	1			100% (1)
Codeine				
Colchicina	1	100% (1)		

Continued on the next page.

Medication	Responses (52)	Yes	No	Undecided
Colchicine (colchis)	1		100% (1)	
Compazine				
Corticoizi	1	100% (1)		
Darvocet				
eye drops				
Flexeril				
folic acid				
folic acid				
folic acid				
I do not know type…				
iron tablets	1		100% (1)	
Keppra				
Keppra				
Lexapro, Zoloft, Effexor…				
Liquid silicone injections				
Midrin as needed				
Migratin?				
Moldamin 1200000 UI	1	100% (1)		
MS Contin for pain				
Paracetamol				
Pax (Valium)				
Penicillin	1	100% (1)		
Petadolex (my daughter…				
Piascledine 300	1	100% (1)		
Prednisone one time…				
Prescription Aleve as needed				
Prilosec				
Prilosec				
Riddalin				
Salazaphrine	1		100% (1)	
STEROIDS, BUT…	1		100% (1)	
Tamal				
various pain relievers	1	100% (1)		
Vicodin				

Continued on the next page.

Medication	Responses (52)	Yes	No	Undecided
vitamin C intravenous therapy	1	100% (1)		
vitamin D				
vitamin D & calcium				
Vitix	1	100% (1)		
Voltarin				
was on Prednisone…	1		100% (1)	
Xanax for the neuropathy				
Zantac				
Zantac				

Number responding = 20 / percent responding = 14% of total respondents (143).
Note: This was a multiple choice question.

Page 87. V. Treatment for Rombergs cont.: Medications Used (page 12 of 13)

1. Please rate how effective this medication is or was:

Medication	Responses (90)	No Change	Some Improvement	Good Improvement	Great Improvement	Total Improvement
Aspirin	6	100% (6)				
Diprosone	2	50% (1)	50% (1)			
Doryx	1	100% (1)				
Dovonex	3	66% (2)	33% (1)			
Doxycycline	2	50% (1)	50% (1)			
Gabapentin	2	100% (2)				
Intravenous Gamma Globulin (IVIG)	2	100% (2)				
Methotrexate	14	64% (9)	21% (3)		14% (2)	
Methylprednisolone	2	100% (2)				
Monocycline	1	100% (1)				
Oral Calcitriol	2	50% (1)	50% (1)			
Penicillamine	1	100% (1)				
Plaquenil	3	100% (3)				
Prednisolone	6	33% (2)	33% (2)		33% (2)	
Prednisone	8	87% (7)			12% (1)	
Quinacrine	2	100% (2)				
Topical antibiotics	2	100% (2)				
vitamin B-12	3	66% (2)	33% (1)			
List of "Other Medications" Follows						
2.9 gram Rocephin...						
Adderall	1			100% (1)		
Albuterol						
Albuterol						
anti seizure meds						
anti seizure meds						
anti seizure meds						
anti seizure meds						
anti seizure meds						
Ceftriaxone						

Continued on the next page.

Medication	Responses (90)	No Change	Some Improvement	Good Improvement	Great Improvement	Total Improvement
Clobetasol Propionate cream	1	100% (1)				
Codeine	1		100% (1)			
Colchicina	1	100% (1)				
Colchicine (colchis)	1	100% (1)				
Compazine	1			100% (1)		
Corticoizi	1	100% (1)				
Darvocet	1		100% (1)			
eye drops	1				100% (1)	
Flexeril	1		100% (1)			
folic acid	1	100% (1)				
folic acid						
folic acid						
I do not know type…						
iron tablets	1	100% (1)				
Keppra						
Keppra						
Lexapro, Zoloft, Effexor…						
Liquid silicone injections						
Midrin as needed						
Migratin?						
Moldamin 1200000 UI	1	100% (1)				
MS Contin for pain	1		100% (1)			
Paracetamol						
Pax (Valium)						
Penicillin	1	100% (1)				
Petadolex (my daughter…	1				100% (1)	
Piascledine 300	1	100% (1)				
Prednisone one time…						
Prescription Aleve as needed						

Continued on the next page.

Medication	Responses (90)	No Change	Some Improvement	Good Improvement	Great Improvement	Total Improvement
Prilosec						
Prilosec						
Riddalin	1				100% (1)	
Salazaphrine	1	100% (1)				
STEROIDS, BUT…	1	100% (1)				
Tamal						
various pain relievers	1			100% (1)		
Vicodin	1			100% (1)		
vitamin C intravenous therapy	1		100% (1)			
vitamin D	1		100% (1)			
vitamin D & calcium	1	100% (1)				
Vitix	1	100% (1)				
Voltarin						
was on Prednisone…	1	100% (1)				
Xanax for the neuropathy	1		100% (1)			
Zantac						
Zantac						

Number responding = 30 / percent responding = 21% of total respondents (143).
Note: This was a multiple choice question.

Page 88. V. Treatment for Rombergs cont.: Medications Used (page 13 of 13)

1. Do you feel taking this medication was worth the effort?

Medication	Responses (91)	Yes	No	Undecided
Aspirin	4	25% (1)	25% (1)	50% (2)
Diprosone	2	50% (1)		50% (1)
Doryx	1			100% (1)
Dovonex	2	50% (1)		50% (1)
Doxycycline	2	50% (1)		50% (1)
Gabapentin	2	50% (1)		50% (1)
Intravenous Gamma Globulin (IVIG)	2			100% (2)
Methotrexate	17	47% (8)	11% (2)	41% (7)
Methylprednisolone	3	66% (2)		33% (1)
Monocycline	1			100% (1)
Oral Calcitriol	1	100% (1)		
Penicillamine	1			100% (1)
Plaquenil	2	50% (1)	50% (1)	
Prednisolone	5	60% (3)		40% (2)
Prednisone	9	44% (4)	33% (3)	22% (2)
Quinacrine	1		100% (1)	
Topical antibiotics	1		100% (1)	
vitamin B-12	2	100% (2)		
List of "Other Medications" Follows				
2.9 gram Rocephin...	1	100% (1)		
Adderall	1	100% (1)		
Albuterol				
Albuterol				
anti seizure meds				
anti seizure meds				
anti seizure meds				
anti seizure meds				
anti seizure meds				
Ceftriaxone				
Clobetasol Propionate cream	1		100% (1)	
Codeine	1	100% (1)		
Colchicina	1		100% (1)	

Continued on the next page.

Medication	Responses (91)	Yes	No	Undecided
Colchicine (colchis)	1			100% (1)
Compazine	1	100% (1)		
Corticoizi	1		100% (1)	
Darvocet	1	100% (1)		
eye drops	1	100% (1)		
Flexeril	1	100% (1)		
folic acid	1			100% (1)
folic acid				
folic acid				
I do not know type…				
iron tablets	1		100% (1)	
Keppra				
Keppra				
Lexapro, Zoloft, Effexor…				
Liquid silicone injections				
Midrin as needed				
Migratin?				
Moldamin 1200000 UI	1		100% (1)	
MS Contin for pain	1	100% (1)		
Paracetamol	1			100% (1)
Pax (Valium)	1			100% (1)
Penicillin	1	100% (1)		
Petadolex (my daughter…	1	100% (1)		
Piascledine 300	1		100% (1)	
Prednisone one time…				
Prescription Aleve as needed				
Prilosec				
Prilosec				
Riddalin	1	100% (1)		
Salazaphrine	1		100% (1)	
STEROIDS, BUT…	1		100% (1)	
Tamal	1			100% (1)
various pain relievers	1	100% (1)		
Vicodin	1	100% (1)		

Continued on the next page.

Medication	Responses (91)	Yes	No	Undecided
vitamin C intravenous therapy	1	100% (1)		
vitamin D	1	100% (1)		
vitamin D & calcium	1			100% (1)
Vitix	1		100% (1)	
Voltarin	1			100% (1)
was on Prednisone…	1		100% (1)	
Xanax for the neuropathy	1	100% (1)		
Zantac				
Zantac				

Number responding = 35 / percent responding = 24% of total respondents (143).
Note: This was a multiple choice question.

Page 88. V. Treatment for Rombergs cont.: Medications Used (page 13 of 13)
2. Comment on medications used:

Responses (18)
Adderall used to combat fatigue. Compazine used to combat nausea.
I have not had medication.
They were expensive, had side effects and there weren't any improvements. I look worse than before the treatment.
Darvocet and Vicodin for pain can take the hard edge off the pain, but not eliminate it. Aspirin and Tylenol are useless with Romberg pain. I'm allergic to Motrin and Codeine, so don't have an opinion on the effectiveness of those for pain management.
It is very well documented that vitamin D, or lack of, plays a significant role in the autoimmune process. I feel that it is definitely worth the effort. It is inexpensive and easy to find. I very much feel that the Petadolex has changed my daughter's life. I felt that we were losing her to the pain of migraines and that the pain was sucking away her wonderful personality. It is not cheap, but still very much worth the effort and money, and it is an excellent alternative to some of the prescription migraine drugs that she had tried.
It's so hard to measure effectiveness of medications with this disease. No way to tell if it would be worse if we didn't try meds.
I think it was worth trying treatments because you never know if it will work. The Methylprednisolone was very hard on the body but I had to try something. I don't think the Methotrexate is working since the Rombergs continues to progress but the doctor (and my husband) wonder if the Methotrexate is working to some degree. Maybe the progression would have been worse if I wasn't taking it. That is why I have continued to take it.
These medications have helped with my pain.
I can't say the disease stopped (atrophy) because of the drugs. I was on Methotrexate and Colchis at the same time so I can't say which of them worked, or both, or none of them... the atrophy apparently ceased (but I also had PMMA grafts (poly methyl methacrylate)) that could have made it to stop), however now I have facial pain (maybe due to the PMMA!). But I found out acupuncture that is working for the pain and hopefully to prevent ongoing atrophy.
At no time was any medication used for PRS only for Ulcerative Colitus. No doctor prescribed anything for PRS except surgery to remove 'core'.
Still in process of treatment, so it is hard to judge at this time - she has only started treatment December 2008.
Also used UVA photo chemotherapy which put disease on hold for a year.
My Dr. told me to take a baby aspirin every day. I don't take it for my PR.
Would do the same again for sure.
Five months after beginning of medication with Methotrexate, we could stop treatment with eye drops containing Cortisone. To treat the eye with Cortisone for long time will result in cataract.
Yes, but keep in mind that I've only ever taken pain relief to treat symptoms and nothing to treat the disease as such.
I never really felt that these meds helped in any way, yet none of the medical personnel seemed open to my suggestions for alternative therapy, omega 3's, high protein diet, among other suggestions. Also when I suggested Methotrexate for the lesions, they felt it was too late to try to use that as her system was too fragile at this point. She ran out of hospital days after being hospitalized for almost a year, so they shipped her to a nursing home, where she ended up with a huge decubitus ulcer and she eventually died at the young age of 39.
This was awful watching my child double her weight in just a couple months. It was terrible. She had trouble breathing, and devloped asthma, arthritis

Number responding = 18 / percent responding = 13% of total respondents (143).

Page 89. V. Treatment for Rombergs cont.: Other Types of Treatments (page 1 of 3)

1. Select any treatments used:

Treatments	Responses (21)	% of Number responding
Acupuncture	5	33%
Hormones	0	0%
Light therapy	2	13%
Massage	8	53%
Naturopathic medicine	5	33%
Topical vitamin D	1	7%

Number responding = 15 / percent responding = 10% of total respondents (143).
Note: This was a multiple choice question.

Page 89. V. Treatment for Rombergs cont.: Other Types of Treatments (page 1 of 3)

2. For "Other" treatments, please enter:

Other Treatments	Responses (20)	% of Number responding
Chiropractic Therapy	1	🟥 7%
Large doses of Omega 3's	1	🟦 7%
Calendula Oil	1	🟥 7%
Keeping vitamins and minerals in the body	1	🟦 7%
received collagen injections when I was a child	1	🟥 7%
botox needle	1	🟦 7%
Frequency Stimulation	1	🟥 7%
silicone liquid injections into the affected areas.	1	🟦 7%
Bio Alcamid injection	1	🟥 7%
N-Acetyl Cysteine (strong antioxidant)	1	🟥 7%
fish oil - high dose 10 - 15 mLs per day	1	🟦 7%
taking herbal anti-inflammatories	1	🟦 7%
on anti-inflammation diet	1	🟥 7%
I will also try facial gym	1	🟦 7%
PMMA (Dermal Filler) injections	1	🟥 7%
creams to apply to burn skin down initially	1	🟦 7%
homeopathic treatment	1	🟦 7%
Cosmetic - Bio-Alcamid	1	🟥 7%

Continued on the next page.

fatty grafts implanted	1	🟥 7%
silicone injections	1	🟦 7%
Other Treatments	**Responses (20)**	**% of Number responding**

Number responding = 14 / percent responding = 10% of total respondents (143).
Note: This was a multiple choice question.

Note: For the rest of this Treatment section, only the first few words of the "Other Treatments" will be shown.

Page 90. V. Treatment for Rombergs cont.: Other Types of Treatments (page 2 of 3)

1. How long was this treatment used:

Medication	Responses (34)	How Long Was This Treatment Used
Acupuncture	5	6 months so far - 7 months 6 months for about 1 year one year with good results (the pain is over)
Hormones		
Light therapy	2	6 months, 3 times a week for 2 months
Massage	7	3 months Only a few weeks with a massage therapist, now I massage myself 1 1/2 years 8 years 12 months several months so far - 7 months
Naturopathic medicine	4	since December 2008 8 years Since diagnosis 5 months ago. Still using it. four months
Topical vitamin D	1	1 year
List of "Other Treatments" Follows		
Chiropractic….		
Large doses of….	1	12 months
Calendula Oil	1	2 weeks
Keeping vita….	1	keep up daily
received col….	1	injections from age 7-15, 6 times a year or more
botox needle	1	current
Frequency….	1	2 times
silicone liquid….	1	about 2 years
Bio Alcamid….		
N-Acetyl Cys….	1	3 months
fish oil - high….	1	2 years
taking herb….		
on anti-inflamm….		
I will also try….	1	facial gym - beginning now, very enthusiastic about it
PMMA (Derm….	1	PMMA injections - the appearance is better, maybe it prevents future atrophy

Continued on the next page.

creams to apply….	1	a year
homeopathic….		
Cosmetic - B….	1	Bio-Alcamid - 2 times injected
fatty grafts….	1	fatty grafts - lasted for about 10 years
silicone injections	1	silcone injections - lasted about 25 yrs
Medication	**Responses (34)**	**How Long Was This Treatment Used**

Number responding = 20 / percent responding = 14% of total respondents (143).
Note: This was a multiple choice question.

Page 90. V. Treatment for Rombergs cont.: Other Types of Treatments (page 2 of 3)

2. Please list any side effects with this treatment:

Medication	**Responses (21)**	**Side Effects With This Treatment**
Acupuncture	4	none, none, none, none
Hormones		
Light therapy	1	none
Massage	3	none, none, none
Naturopathic medicine	2	Major improvement in overall health. Positive side affects. He has grown in height and weight. No colds or flu.
Topical vitamin D		
List of "Other Treatments" Follows		
Chiropractic....		
Large doses of....		
Calendula Oil	1	none
Keeping vita....	1	seems to slow the progression
received col....	1	none-collagen would reabsorb within a week
botox needle		
Frequency....	1	None.. and no apparent benefit as of yet.
silicone liquid....	1	Silicone moved from under the tight skinned Romberg's areas into normal areas resulting in hard lumps which made the difference between the non-fat Romberg's areas & the now silicone areas far more noticable, had to have an operation to try to drain the silicone out but this did not work.
Bio Alcamid....	1	infection
N-Acetyl Cys....		
fish oil - high....		
taking herb....		
on anti-inflamm....		
I will also try....	1	none
PMMA (Derm....	1	PMMA - possible pain? not sure if the pain is from PRS or from PMMA
creams to apply....	1	none
homeopathic....		
Cosmetic - B....	1	none
fatty grafts....	1	fatty graft. - one was infected and needed to be replaced
silicone injections		

Number responding = 14 / percent responding = 10% of total respondents (143).
Note: This was a multiple choice question.

Page 91. V. Treatment for Rombergs cont.: Other Types of Treatments (page 3 of 3)

1. When treatment ended, did the symptoms reappear after a period of time:

Treatment	Responses (27)	Yes	No	Undecided
Acupuncture	3	66% (2)	33% (1)	
Hormones				
Light therapy	2	100% (2)		
Massage	6	42% (3)	14% (1)	28% (2)
Naturopathic medicine	2			100% (2)
Topical vitamin D				
List of "Other Treatments" Follows				
Chiropractic….				
Large doses of….	1	100% (1)		
Calendula Oil				
Keeping vita….	1			100% (1)
received col….	1	100% (1)		
botox needle				
Frequency….	1			100% (1)
silicone liquid….	1	100% (1)		
Bio Alcamid….	1	100% (1)		
N-Acetyl Cys….	1			100% (1)
fish oil - high….	1			100% (1)
taking herb….				
on anti-inflamm….				
I will also try….	1		100% (1)	
PMMA (Derm….	1		100% (1)	
creams to apply….	1		100% (1)	
homeopathic….				
Cosmetic - B….	1	100% (1)		
fatty grafts….	1		100% (1)	
silicone injections	1		100% (1)	

Number responding = 17 / percent responding = 12% of total respondents (143).
Note: This was a multiple choice question.

Page 91. V. Treatment for Rombergs cont.: Other Types of Treatments (page 3 of 3)

2. Please rate how effective you think this treatment was:

Medication	Responses (32)	No Change	Some Improvement	Good Improvement	Great Improvement	Total Improvement
Acupuncture	5		60% (3)	20% (1)	20% (1)	
Hormones						
Light therapy	2	50% (1)	50% (1)			
Massage	7	25% (2)	25% (2)		37% (3)	
Naturopathic medicine	3		66% (2)		33% (1)	
Topical vitamin D						
List of "Other Medications" Follows						
Chiropractic….						
Large doses of….	1				100% (1)	
Calendula Oil						
Keeping vita….	1		100% (1)			
received col….	1	100% (1)				
botox needle	1				100% (1)	
Frequency….	1	100% (1)				
silicone liquid….	1	100% (1)				
Bio Alcamid….	1			100% (1)		
N-Acetyl Cys….	1		100% (1)			
fish oil - high….	1		100% (1)			
taking herb….						
on anti-inflamm….						
I will also try….	1		100% (1)			
PMMA (Derm….	1		100% (1)			
creams to apply….	1					100% (1)
homeopathic….						
Cosmetic - B….	1			100% (1)		
fatty grafts….	1				100% (1)	
silicone injections	1					100% (1)

Number responding = 20 / percent responding = 14% of total respondents (143).
Note: This was a multiple choice question.

Page 91. V. Treatment for Rombergs cont.: Other Types of Treatments (page 3 of 3)

3. Comments for Other Types of Treatments:

Responses (12)
Chiropractic treatment and massage therapy along with high doses of Omega 3's.
While painfully slow, we believe we may have found a treatment that works in reversal of some symptoms.
Had my own body fat injected by a plastic surgeon under affected areas. Was reabsorbed in the scalp but some hair started to grow - some stayed in the forehead. Mild success.
Silicone injections made my face worse.
The appearance was good / but my body rejected the material (Bio-Alcamid).
He was diagnosed just four months ago. Been on the naturopathic treatment for the whole time. Atrophy may have slowed down, hard to tell. But is gaining weight, growing in height and has so far avoided cold season entirely.
I developed a depression in my cheek area about a month after stopping the UVA treatment. It makes me wonder whether the UVA sped it up or if it was controlling the Rombergs, and stopping the UVA made the progression speed up. No one can tell me that.
Anti-oxidants and fish oil - no improvement in symptoms seen, I am just aiming to slow the progression. I think the evidence backing these treatments in a variety of inflammatory and/or degenerative conditions is very positive and I am hopeful they can work just as well in PRS.
Hard to tell, it has only been 2 months.
We are still trying with homeopathic treatment and some changes are there...
The massage helped the face feel more refreshed and 'lighter'. It must have helped with blood circulation. I don't know if it would help to stop the atrophy or help plump up the face again.
The naturopathic medicine is hit and miss until we can figure out the right combination. We believe it is slowing the atrophy. It is helping his general health. He really hadn't grown much in the last two years. Now he is growing in both weight and height. Has not caught any of the colds or flu that his siblings have brought home. His overall health has greatly improved.

Number responding = 12 / percent responding = 8% of total respondents (143).

Page 92. V. Treatment for Rombergs cont.: Surgery (page 1 of 7)

1. Please select the type of surgery that the affected person has had:

Type of Surgery	Responses (146)	% of Number responding
Artificial implants	9	16%
Bone implant	7	12%
Eye	11	19%
Fat graft	21	36%
Fat injections	25	43%
Free flap or flap procedure	9	16%
Free flap or flap procedure with microvascular surgery	21	36%
Injection of artificial material	11	19%
Jaw reconstruction	6	10%
Nose	12	21%
Revision surgery	14	24%

Number responding = 58 / percent responding = 41% of total respondents (143).
Note: This was a multiple choice question.

Page 92. V. Treatment for Rombergs cont.: Surgery (page 1 of 7)

2. For "Other" surgeries, please enter:

Other Surgeries	Responses (37)	% of Number responding
artificial implant on right upper eyebrow--orbit	1	🟥 5%
eyelid surgergy to correct previous surgery	1	🟦 5%
lower implant of orbit and temple	1	🟥 5%
fat injections	1	🟦 5%
reconstructive-free flap procedure	1	🟥 5%
forehead	1	🟦 5%
Small multiple Reconstructive Facial Surgeries	1	🟥 5%
Many surgeries	1	🟦 5%
SILICONE IMPLANTED IN MY FOREHEAD	1	🟥 5%
Revision & fat injection	1	🟦 5%
Free flap microvascular surgery - implant on cheek bone	1	🟥 5%
Revision & fat injection	1	🟦 5%
Sculptra	1	🟥 5%
botox injections to ease the double vision	1	🟦 5%
liposuction to reduce the muscle after free flap	1	🟥 5%
Free flap	1	🟦 5%
Fat graft	1	🟥 5%
my surgery	1	🟦 5%
a smaller surgery on my upper lip when I was a child	1	🟥 5%

Continued on the next page.

Other Surgeries	Responses (37)	% of Number responding
debulking graft	1	🟦 5%
lifting graft	1	🟥 5%
cartilage taken from under the right breast and implanted in the right cheek	1	🟦 5%
My first surgery	1	🟥 5%
My second surgery (eye brow)	1	🟦 5%
My third was fat injections	1	🟥 5%
trapano parietal posterior izquierdo at age 3	1	🟦 5%
Remove some dying tissue on his face	1	🟥 5%
Repair donor site on his belly after Mitch tore it open	1	🟦 5%
bone graft and dental implants	1	🟥 5%
Dr. rebuilt eye socket using bone grafts from my scalp	1	🟦 5%
lower left eyelid was revised using cartilege from my right ear	1	🟥 5%
bald spot on En coup de sabre was excised and edges sewn together	1	🟦 5%
Dr. pulled eyebrow together and sewed it in hopes of getting rid of bald spot in it	1	🟥 5%
She has had only one surgery in 2001.	1	🟦 5%
laser treatment for discoloration (did not work)	1	🟥 5%

Continued on the next page.

leather skin transplant	1	🟦 5%
brain surgery for removal of lesions	1	🟥 5%
Other Surgeries	**Responses (37)**	**% of Number responding**

Number responding = 21 / percent responding = 15% of total respondents (143).
Note: This was a multiple choice question.

Note: For the rest of this Surgery section, only the first few words of the "Other Surgeries" will be shown.

Page 93. V. Treatment for Rombergs cont.: Surgery (page 2 of 7)

1. Please select the number of surgeries:

Surgery	Responses (174)	1	2	3	4	5	6	7	8	9	10	More than 10
Artificial implants	10	63% (7)	18% (2)		9% (1)							
Bone implant	7	85% (6)	14% (1)									
Eye	11	45% (5)	9% (1)		36% (4)			9% (1)				
Fat graft	21	52% (11)	33% (7)	4% (1)	4% (1)	4% (1)						
Fat injections	26	34% (9)	42% (11)	15% (4)	7% (2)							
Free flap or flap procedure	9	37% (6)	18% (3)									
Free flap or flap procedure with microvascular surgery	22	72% (16)	13% (3)		9% (2)	4% (1)						
Injection of artificial material	13	7% (1)	15% (2)	7% (1)	15% (2)	15% (2)		15% (2)				23% (3)
Jaw reconstruction	6	50% (3)	50% (3)									
Nose	12	83% (10)	8% (1)	8% (1)								
Revision surgery	17	41% (7)	23% (4)	29% (5)		5% (1)						
List of "Other Surgeries" Follows												
artificial implant on....												
eyelid surgergy to....												
lower implant of o....												
fat injections												
reconstructive-fre....												
forehead	1	100% (1)										
Small multiple R....	1										100% (1)	
Many surgeries	1	100% (1)										

Continued on the next page.

Surgery	Responses (174)	1	2	3	4	5	6	7	8	9	10	More than 10
SILICONE IMP….	1	100% (1)										
Revision & fat…2												
Free flap micro….												
Revision & fat…1												
Sculptra	1		100% (1)									
botox injections….												
liposuction to….												
Free flap												
Fat graft												
my surgery	1	100% (1)										
a smaller su….	1	100% (1)										
debulking graft	1	100% (1)										
lifting graft	1	100% (1)										
cartilage taken….	1		100% (1)									
My first surgery												
My second su….												
My third was fat….												
trapano parietal….	1	100% (1)										
Remove some….	1	100% (1)										
Repair donor….	1	100% (1)										
bone graft and….	1	100% (1)										
Dr. rebuilt eye….	1	100% (1)										
lower left eyelid….	1	100% (1)										
bald spot on En….	1	100% (1)										
Dr. pulled eye….												

Continued on the next page.

She has had o….												
laser treatment….	1	100% (1)										
leather skin tra….	1	100% (1)										
brain surgery….	1			100% (1)								
Surgery	**Responses (174)**	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**More than 10**

Number responding = 60 / percent responding = 42% of total respondents (143).
Note: This was a multiple choice question.

Page 94. V. Treatment for Rombergs cont.: Surgery (page 3 of 7)

1. Please enter the number of surgeries if the number of surgeries is more than 10:

Surgery	Responses (18)	Number of Surgeries Is More Than 10
Artificial implants	2	1, 4
Bone implant	1	to straighten my arch of upper teeth
Eye	1	lifting lower lid
Fat graft	1	1
Fat injections	3	15, 2, 2
Free flap or flap procedure	1	1
Free flap or flap procedure with microvascular surgery	4	1, 1, 1, 1
Injection of artificial material	3	Straight silicone injections over a period of 3 years Several tiny injections are done to fill the indentation or correct asymmetry. no idea - went on for years as a child
Jaw reconstruction		
Nose		
Revision surgery	1	debulking graft
List of "Other Surgeries" Follows		
artificial implant on….		
eyelid surgergy to….		
lower implant of o….		
fat injections		
reconstructive-fre….		
forehead		
Small multiple R….		
Many surgeries		
SILICONE IMP….		
Revision & fat…2		
Free flap micro….		
Revision & fat…1		
Sculptra		
botox injections….		
liposuction to….		
Free flap		
Fat graft		

Continued on the next page.

Surgery	Responses (18)	Number of Surgeries Is More Than 10
my surgery		
a smaller su….		
debulking graft	1	lifting graft
lifting graft		
cartilage taken….		
My first surgery		
My second su….		
My third was fat….		
trapano parietal….		
Remove some….		
Repair donor….		
bone graft and….		
Dr. rebuilt eye….		
lower left eyelid….		
bald spot on En….		
Dr. pulled eye….		
She has had o….		
laser treatment….		
leather skin tra….		
brain surgery….		

Number responding = 9 / percent responding = 6% of total respondents (143).
Note: This was a multiple choice question.

Page 95. V. Treatment for Rombergs cont.: Surgery (page 4 of 7)

1. Please enter the age of the person when this surgery was performed:

Surgery	Responses (136)	Age When This Surgery Was Performed
Artificial implants	10	Just under 18, 32 & 33, 14, 17 years old, 16 - 20, 17, 22, 48, 15, 24
Bone implant	6	17, 15, 17, 12, 22, 13 and 16
Eye	9	17, 17, 15 - 18 - 22, 52, 51, Ages varied from 25 - 65, 50 - 51 - 52, 30, 38 and 39
Fat graft	14	30, Just under 18, 38, 24, 11, 20, 17, 12 and 15, 9, 21, 29, (36, 37, 38, 39, 40), (14, 15, 16), 21
Fat injections	21	56 and 57, 30, 17, 15 and 16, 33 and 34, 32 and 33, 28, 32, 16, 9 and 10, (24, 25, 26), First: 70 and Second: 70, 49 and 49, 22 and 32, (10, 11, 14, 20), 29, 21, 28 and 30, 49 and 50, 15.5 and 16.5, 34
Free flap or flap procedure	7	17, 14, 12 - 17, 19, 13 and 15, 30, 21
Free flap or flap procedure with microvascular surgery	18	28, 16, 15 and 16, 16, 36, 24 or 25, 12, 45, 23, 41, 13, 34, 32, 12, (37, 38, 39, 40), 48, 23, 15
Injection of artificial material	8	(18, 19, 20, 21, 22, 23, 24, 25, 26, 27, 28, 29), (36, 37, 38, 39), 13 - 16, (22, 23, 24), 14, collagen, 24, 14 and 15
Jaw reconstruction	4	15, 14, 17, 13 and 16
Nose	8	22, 15, 16, 18, 19, (14, 16, 48), 28, 32
Revision surgery	13	18, 37, 25 and 26, 12, 22 and 32, 22 - 24, 18, 13 and 13, (35, 36, 37), (14, 15, 16), 49 and 50, 30, 16.5
List of "Other Surgeries" Follows		
artificial implant on….		
eyelid surgegy to….		
lower implant of o….		
fat injections		
reconstructive-fre….		
forehead	1	5
Small multiple R….	1	23, 24, 25, 26, 27, 28, 30
Many surgeries		
SILICONE IMP….	1	23
Revision & fat…2		
Free flap micro….		
Revision & fat…1		
Sculptra	1	27
botox injections….		

Continued on the next page.

Surgery	Responses (136)	Age When This Surgery Was Performed
liposuction to….		
Free flap		
Fat graft		
my surgery	1	22
a smaller su….	1	11
debulking graft	1	45
lifting graft	1	46
cartilage taken….	1	21
My first surgery		
My second su….		
My third was fat….		
trapano parietal….	1	3
Remove some….	1	12
Repair donor….	1	12
bone graft and….	1	51
Dr. rebuilt eye….	1	33ish
lower left eyelid….	1	34ish
bald spot on En….	1	35ish
Dr. pulled eye….		
She has had o….		
laser treatment….	1	28
leather skin tra….	1	22
brain surgery….		

Number responding = 54 / percent responding = 38% of total respondents (143).
Note: This was a multiple choice question.

Page 96. V. Treatment for Rombergs cont.: Surgery (page 5 of 7)
1. Please provide a brief description of the surgery:

Surgery	Responses (117)	Description of the Surgery
Artificial implants	10	small cheek implant Upper cheek. Artificial material is H something something. (three letters - I forget exactly) Implanted titanium into the chin area and injected fat into the cheek. Plastic cheek bone implanted took tissue from my breast to implant in face... tissue rejected 2 attempts, both taken out after repulsion (?) a silicone implant was placed in my forehead Methylmethacrolate to forehead and temple Small implant during initial free flap upper eyebrow - orbit of eye
Bone implant	3	bone graft from left hip to right forehead bone from skull transfered to forehead and cheek bone Moved my cheek bone to where it should be and implanted bone behind.
Eye	8	lift eyelid leather skin transplant During strabismus surgery the eyelid has a suture installed to raise the eyelid. lifting lower lid Eye muscle & scleral graft and eyelid lifts. 2 squint to get the eye straight, cataract removed, detached retina was folded over PRK eye Surgery squint surgery and both eyes fixed into position to correct double vision
Fat graft	14	Fat removed from right butt and inserted into right check & jaw through same incision along front of right ear. Fat was taken from my stomach and implanted into my cheek. fat taken from bottom from her stomach. fat graft was taken from my groin area and put in by layers into the cheek fat taken from buttock and inserted into cheek and chin from hip/groin area to lip. Mostly reabsorbed and didn't work fat taken from buttocks area and inserted in cheek from lower abdomen to cheek area and lower forehead left side did not last, fat was reabsorbed within 3 months Fat was taken from the inside of my knees and injected into my face. Need 2 or 3 more procedures depending on how much fat stays. Improvement can be seen. thigh and groin fat used to reconstruct cheek, lip and temple Fat from groin area placed in chin, tissue from inside cheek placed in lip taken from hip and grafted to side

Continued on the next page.

Surgery	Responses (117)	Description of the Surgery
Fat injections	17	fat injections to left and right face and chin 2 Fat injections, one month apart, fat was reabsorbed but color of skin better. fat liposuction taken from thighs then put all over my face in small amounts butt tissue injected into jaw dissappeared after a few weeks fat transfer from stomach to cheek removed from my stomach and injected into the cheek twice. Reabosrbed once and infected the second time. Dr. harvested from my tummy, spun it in centrifuge, injected it- painful!!! fat transer to left cheek using fat from my abdomen Fat liposucted from knees and injected in cheek and lips. Completely reabsorbed, didn't work. June 2008: Hospital under general anesthesia. Fat from stomach was injected into right side of face.... side of face, cheek and chin. Second set in January 2009. In-office procedure...fat injected into side of face in area in which fat from previous procedure had been reabsorbed. Fat was taken from various parts of the body and injected into various parts of the face (after it was spun out etc) tissue from abdomen x2 forehead and cheek in scalp, forehead and nose to remove excess tissue and redistribute transferred tissue taken from abdominal and placed in cheek and upper lip area
Free flap or flap procedure	4	done at age 17 first surgery removal of area on cheek and grafted to nose they shaved part of my skull from the back and brought it to my forehead taken from my head to my chin
Free flap or flap procedure with microvascular surgery (Continued next page)	17	Galial scalp flap: Section of scalp inserted into right check & jaw through incision along front of right ear. silicone was removed and 4 fatty grafts were implanted Blood vessels removed and implanted in face (taken from back of shoulder down around the under arm area). tissue taken from shoulder flap taken from groin and inserted into cheek, top lip and chin. supplied with blood from vein in neck flap from scapular area- hooked up blood vessels, etc., used bolsters to hold flap in place tissue transferred from groin after soft tissue was implanted, free flap was used to fill in space free flap from my upper leg (sorry for my English) flap from shoulder blade area took muscle from my chest and skin from my leg and transplanted into my face

Continued on the next page.

Surgery	Responses (117)	Description of the Surgery
(Continued from previous page) Free flap or flap procedure with microvascular surgery	17	A flap was removed from my left thigh to my face. Done 12/15/2008 with Dr Siebert, New York University; flap from back implanted right side of face/cheek. Took shoulder muscle to implant into the cheek, Surgeon put too much in and I had to have 3 more surgeries to remove excess tissue tissue taken from uppper right thigh and transferred into right cheek and chin Surgery on left side of face major initial surgery - tissue taken from shoulder area
Injection of artificial material	10	Restylane stays, but get clogged and moves around Sculptra on forehead and cheek - 2 sessions Radiesse injections Bio Alcamid has lasted, Radiance dissolved within 3 months injections of PMMA (poly methyl methacrolate) in different areas: chin, cheek, under eyes, temple, with revisions When I was 14 years old I was given 'liquid silicone injections' by a doctor who was licensed by the government to experiment with this procedure on people with facial abnormalities. With this procedure liquid silicone is injected directly into the tissue to fill out facial indentations caused by the Rombergs. injections from age 7-15, 6 times a year or more to forehead and nose Reconstructive surgery Liquid silicone, head, chin
Jaw reconstruction	3	jaw straighten removal of affected area on lip and gum, removal of teeth Leforte osteotomy, sagital split osteotomy with plates and screws
Nose	6	part of lobe from left ear to enlarge right nostril nose straighten tried to make nose more symmetrical removal of area in one cheek and grafted to nose a small section of my right nostril was removed to make it smaller and then added to the left side to make it bigger implant
Revision surgery	11	one fatty graft was infected and replaced tissue taken out because of overfill excess bulk from flap liposuctioned had to de-bulk the flap like 3 times to contour it to get it right... debulking graft more fat grafts and injections done tidy up of lip scarring debulking the graft to remove excess tissue and redistribute transferred tissue PRK (photorefractive keratectomy) eye surgery 6 months post initial surgery & another revision 2 years later

Continued on the next page

List of "Other Surgeries" Follows		
Surgery	**Responses (117)**	**Description of the Surgery**
artificial implant on….		
eyelid surgergy to….		
lower implant of o….		
fat injections		
reconstructive-fre….		
forehead	1	Cutting away of discolored En coup de sabre mark on forehead
Small multiple R….	1	Cutting away of discolored En coup de sabre mark on forehead
Many surgeries		
SILICONE IMP….	1	SEE PRIOR ANSWER
Revision & fat…2		
Free flap micro….		
Revision & fat…1		
Sculptra		
botox injections….		
liposuction to….		
Free flap		
Fat graft		
my surgery	1	"z" incision on chin to relieve skin tightness
a smaller su….	1	A small piece of tissue was removed from + to my upper lip
debulking graft	1	lifting graft
lifting graft		
cartilage taken….	1	cartilage from under right breast put in right cheek
My first surgery		
My second su….		
My third was fat….		
trapano parietal….	1	"tarpano parietal posterior izquierda"
Remove some….		
Repair donor….		
bone graft and….	1	Bone harvested from good side of jaw and put into affected side. When it had taken, I was able to have dental implants put in.
Dr. rebuilt eye….	1	Dr. excised bald spot from en coup de sabre, and sewed bald spot together in eyebrow
lower left eyelid….	1	used bone grafts to rebuild eye socket on PRS side

Continued on the next page.

bald spot on En….	1	Used cartilege from right ear to rebuild lower left PRS eyelid which had issues
Dr. pulled eye….		
She has had o….		
laser treatment….	1	did not work
leather skin tra….	1	leather skin transplant
brain surgery….		
Surgery	**Responses (117)**	**Description of the Surgery**

Number responding = 50 / percent responding = 35% of total respondents (143).
Note: This was a multiple choice question.

Page 97. V. Treatment for Rombergs cont.: Surgery (page 6 of 7)

1. Please rate the outcome of this surgery:

Surgery	Responses (148)	No Change	Some Improvement	Good Improvement	Great Improvement	Total Improvement
Artificial implants	10	20% (2)	10% (1)	50% (5)	10% (1)	10% (1)
Bone implant	6		50% (3)	16% (1)	16% (1)	16% (1)
Eye	10		40% (4)	20% (2)	30% (3)	10% (1)
Fat graft	17	11% (2)	38% (7)	22% (4)	22% (4)	
Fat injections	24	12% (3)	29% (7)	33% (8)	8% (2)	16% (4)
Free flap or flap procedure	8	13% (1)	13% (1)	75% (6)		
Free flap or flap procedure with microvascular surgery	19	5% (1)	10% (2)	15% (3)	57% (11)	10% (2)
Injection of artificial material	10	20% (2)	40% (4)		30% (3)	10% (1)
Jaw reconstruction	4		50% (2)		50% (2)	
Nose	10		60% (6)	30% (3)	10% (1)	
Revision surgery	15		26% (4)	13% (2)	40% (6)	20% (3)
List of "Other Surgeries" Follows						
artificial implant on….						
eyelid surgergy to….						
lower implant of o….						
fat injections						
reconstructive-fre….						
forehead	1	100% (1)				
Small multiple R….	1					100% (1)
Many surgeries						
SILICONE IMP….						
Revision & fat…2						
Free flap micro….						
Revision & fat…1						
Sculptra						
botox injections….						
liposuction to….						

Continued on the next page.

Surgery	Responses (148)	No Change	Some Improvement	Good Improvement	Great Improvement	Total Improvement
Free flap						
Fat graft						
my surgery	1		100% (1)			
a smaller su….						
debulking graft	1			100% (1)		
lifting graft	1		100% (1)			
cartilage taken….	1		100% (1)			
My first surgery						
My second su….						
My third was fat….						
trapano parietal….	1		&100% (1)			
Remove some….	1					100% (1)
Repair donor….	1					100% (1)
bone graft and….	1					100% (1)
Dr. rebuilt eye….	1				100% (1)	
lower left eyelid….	1				100% (1)	
bald spot on En….	1				100% (1)	
Dr. pulled eye….						
She has had o….						
laser treatment….	1	100% (1)				
leather skin tra….	1		100% (1)			
brain surgery….						

Number responding = 59 / percent responding = 41% of total respondents (143).
Note: This was a multiple choice question.

Page 98. V. Treatment for Rombergs cont.: Surgery (page 7 of 7)

1. Any suggestions that you would like to pass on:

Responses (20)
Begin with fat injections, then proceed to reconstruction.
Seek advice from as many health care professionals who have had experience in dealing with PR . Seek support and advice from people who have experiences themselves. Do not travel this road alone.
Surgery is a lot to go through. Rely on those who love you, to help you get through it.
Before thinking on Surgery, make sure the Rombergs is not active.
Do not get liquid silicone injections. They're dangerous. The material moves and hardens, causing further disfigurement. It is difficult, if not impossible, to remove from the tissue once injected. I have experienced terrible 'infections' (?) around the main site of the liquid silicone injections. This started happening 23 years after the procedure was done. These 'infections' are debilitating when they occur, needing several months to recover. What happens is the area around the silicone begins to swell and there is a localized pain that is different than a 'dent' headache. There is an intense fatigue and sometimes I am nearly unable to move as my body feels as though it is moving through sludge. The strain in my left eye becomes more pronounced causing pain and redness. The joints in my hands, and sometimes my ankles, become inflamed with a searing pain, and I get painful tongue ulcers. When this silicone related 'infection' happens doctors have tried antibiotics, but that has proven unsuccessful as a treatment. What I have learned to do as a "treatment" is when I first start to notice these symptoms begin, I rest a lot and keep myself clear of stress as much as possible. I double - up on multi- vitamins and eat spicy foods (more on that remedy later). This approach seems to help manage the severity and duration of the silicone 'infection' problem.
Since my son still has a 2nd surgery to complete the process, it is hard to determine the final outcome of the surgery at this point. The flap was successfully implanted though he had much swelling after the surgery and still does, after about 10 weeks. It is our hope that this will stop the progression of PRS, it's why we chose this route.
I would suggest waiting to make sure the Rombergs is in remission before doing any surgeries.
The PMMA injections (poly methyl methacrolate) are not recommended by many surgeons. However they are definitive, cheap, they are used for HIV patients with lipodistrophy. There was certain degree of reabsorption but even though, the injections were a great success.
Free flap transplant was hard but worth it. After that it was too much fat but with lipolyse I'm getting close to an amazing result.
Remember this, if people like you, they like for who you are, not what you look like.
Live life to the fullest. There is always someone who is worse off than you.
With every surgery you gain some, yet lose some, or else makes something else worse.
For me, fat injections worked the best.
Do your homework with surgeons!! It's important to chose one who understands and has experience with Parry Romberg Syndrome. If patient is a child, be sure that they understand what the surgery is all about. Give the child choices and time to contemplate. We would recommend that the child be old enough to make their OWN informed decision, unless medically necessary.
Dental implants are expensive but have made such a difference.
Some of the questions were confusing... about en coup de sabre - was the progression question about progression of Rombergs or the en coup de sabre? I might have answered it wrong, sorry!
I can't think of any, but I'm sure that I will after I finish this...
Continued on the next page.

Page 98. V. 1. Responses (continued)
It seems so many things have been done and almost nothing was succesful. But after many surgeries, I felt happy and even pretty enough, but then the symptoms worsened, without any cause.
Since I am older now, if I need to have a fatty graft replaced I am considering some type of injections to be done. The graft surgery is too lengthy and is painful. I just don't know if I can go through another long surgery again.
Do not have surgery until you are sure the Romberg has burnt itself out, which is still a risk, because some cases don't.

Number responding = 20 / percent responding = 14% of total respondents (143).

Page 98. V. Treatment for Rombergs cont.: Surgery (page 7 of 7)

2. Comments for Surgery:

Responses (19)
Fat injections was successful but only for about a year. Free flap was most successful but a long way from perfect. Other surgeries were mostly corrective or to compensate for infections.
DON'T USE SILICONE. IT DOESN'T STAY IN PLACE. IT'S IN MY BODY SOMEWHERE, BUT I DON'T KNOW WHERE.
Initial surgery can be very daunting and a person needs time to heal and recover in every respect. There is never a 'right' time for surgery and it may not be the option for everybody. Difficult decison to make, so best to be very well informed.
Fat injections looked good for about 2 months and then reabsorbed. Cheek implant helps, but I think it's too small. Sculptra - had 2 sessions, and is a gradual acting material. Jury is still out on this one.
Before having surgery, ask as many questions as you think. It's too late after the procedure has been done.
It wasn't easy to watch our son go through such an invasive surgery but the alternative -- watching the atrophy of his face -- was unbearable. So if successful, I would highly recommend this option with Dr. Siebert and his staff at New York University.
My free flap surgery is not perfect but I did not regret anyway. I look prettier than before the surgery.
She will be having a free flap tissue graft in the next six months.
When I was younger, I was all set to do whatever it took to look better, even though it has progressed more now, I am weary about having more surgery, I have seen a plastic surgeon who wants to do several surgeries to help, but am afraid to get them done.
The surgery was very difficult and it took me a long time to recover. I felt it took me 10 full years to really feel like Rombergs wasn't the main focus of my life and that I looked wonderful.
Plastic surgery does not make you look like someone on extreme makeover but very grateful that the surgery seemed to halt progress or else would have eventually affected eye area.
Do your homework.
Satisified with the results.
I believe all the transplants have been reabsorbed.
Know when enough is enough although touch-ups may be necessary. Be realistic about outcome. Don't expect perfect results.
Will have to wait to see how much fat will be reabsorbed.... Quite a bit of fat was reabsorbed from first session of June 2008, and second session was only done in January 2009. Have follow up appointment with surgeon in July 2009. I am very pleased with my progress to date....my face looks much better.
Fat injections are a relatively unobtrusive form of treatment, as the 'filler' is taken from your own body. When using an artificial filler, there is more chance that it can cause problems, be rejected, move around or behave in an unnatural way.
Terrifying, but you have to do it, just in case it makes the difference you long for.
The surgery took at first but then reabsorbed.

Number responding = 19 / percent responding = 13% of total respondents (143).

Page 99. VI. Any Other Affected Family Members

1. Are there any other family members with facial and/or optical asymmetry?

Asymmetry	Responses (125)	% of Number responding
Yes	6	5%
No	119	95%

Number responding = 125 / percent responding = 87% of total respondents (143).

Note: If "Yes" was selected at the above question 99. VI. 1. "Are there any other family members with facial and/or optical asymmetry?" then the following section (Page 100) would apply.

Page 100. VI. Any Other Affected Family Members cont.: Describe Family Member

1. Please provide information about this other family member:

Responses (6)	Relationship	Current Age	Age of onset
1	mother	88	do not know, but have only noticed it in her 80's
1	child	7	? from photos, looks like it started when mine did
1	first cousin	35	
1	daughter	24	first noticed at 19
1	my son	12	maybe 2
1	grandmother	63	don't know she always thought it was normal / thinks it may be from a small stroke that was not diagnosed.

Number responding = 6 / percent responding = 4% of total respondents (143).

Page 100. VI. Any Other Affected Family Members cont.: Describe Family Member

2. Has this person been medically diagnosed with Rombergs?

Diagnosed	Responses (5)	% of Number responding
Yes	0	0%
No	5	100%

Number responding = 5 / percent responding = 3% of total respondents (143).

Note: If "Yes" was selected at the above question 100. VI. 2. "Has this person been medically diagnosed with Rombergs?" then the following section (Page 101) would apply.

Page 101. VI. Any Other Affected Family Members cont.: Diagnostic Information

1. At what age was the affected person diagnosed:

Age	Responses (1)	% of Number responding
13	1	100%

Number responding = 1 / percent responding = 0.6% of the total respondents (143).

Page 101. VI. Any Other Affected Family Members cont.: Diagnostic Information

2. Please select the type of specialist who made the diagnosis:

Specialist	Responses (1)	% of Number responding
Acupuncturist	0	0%
Dentist	0	0%
Dermatologist	0	0%
Ear, Nose and Throat	0	0%
General Practitioner	0	0%
Geneticist	0	0%
Neurologist	0	0%
Opthamologist	0	0%
Plastic Surgeon	1	100%
Reconstructive Surgeon	0	0%
Rheumatologist	0	0%
Other (Specify)	0	0%

Number responding = 1 / percent responding = 0.6% of the total respondents (143).
Note: This was a multiple choice question.

Page 101. VI. Any Other Affected Family Members cont.: Diagnostic Information

3. Please select the procedure used to make the diagnosis:

Procedure	Responses (1)	% of Number responding
ANA (Antinuclear Antibody test for level of inflammation)	0	0%
Biopsy for bacteria	0	0%
Biopsy for sclerotic content	0	0%
CAT (computed axial tomography (CAT or CT scan))	0	0%
MRI (magnetic resonance imaging)	0	0%
Ultrasound	0	0%
Visual Diagnosis	1	100%
Other (Specify)	0	0%

Number responding = 1 / percent responding = 0.6% of the total respondents (143).
Note: This was a multiple choice question.

Page 101. VI. Any Other Affected Family Members cont.: Diagnostic Information

4. Comments for the Affected person's diagnostic information:

Count	Comments (0)

Number responding = 0 / percent responding = 0% of total respondents (143).

Page 100. VI. Any Other Affected Family Members cont.: Describe Family Member

3. Is the person facially affected on:

Facially affected on	Responses (5)	% of Number responding
the Left side	2	40%
the Right side	3	60%
Both sides	0	0%
Not affected	0	0%

Number responding = 5 / percent responding = 3% of total respondents (143).

Page 100. VI. Any Other Affected Family Members cont.: Describe Family Member

4. Where on the affected person's body do the symptoms also appear:

Where on body	Responses (3)	% of Number responding
the Left side	0	0%
the Right side	2	67%
Both sides	0	0%
Not affected	1	33%

Number responding = 3 / percent responding = 2% of total respondents (143).

Page 100. VI. Any Other Affected Family Members cont.: Describe Family Member

5. Comments

Responses (3)
Son is being observed. Chin changes -- rash became white spot, became brown line. Over time, brown line more evident. Initally didn't even notice it, even though can see it in photos from that time on. Now area has depressed a bit. One doctor calling it a lesion to be observed. Other doctor said he thought it was a hybrid LS/PRS, but need to watch it. Very scary and hard to bear. There is no playbook for this. He also has pigment changes body wide and tested positive for a tick bite, although rhemo said didn't have active Lyme? Recently noted "electric shock" on right side of face, which is scary.
My cousin was also born with a squint. I can only notice a slight asymmetry of her face (still probably in the "normal" range). One of her toes is considerably bigger than the other (since puberty), which means she needs 2 different sized shoes. I think these are both on her right side.
Two daughters with early PRS. (3 daughters in all). Neither of the two have a definite diagnosis because we have avoided a diagnosis for insurance purposes. Both have mild symptoms on appearance. Both have right side atrophy. Both have predominantly right side headaches. Both have more apparent atrophy when looking at them in a mirror. The 24 year old's right breast is smaller. She has white matter on her MRI. Her symptoms started at 19. Her facial atrophy remains very mild. Her headaches are less frequent and less severe, but do cause numbness of her extremities at times. She was diagnosed with Juvenile Rheumatoid Arthritis at the age of 3, and has had a multitude of medical issues. The 18 year old has no white matter but her MRI shows asymmetry of her maxillary structures, temporal lobe and sylvian branches. We first noticed her facial asymmetry at the age of 10. It remains very mild. She suffers from twitching on mostly the right side of her body and in her head as well as numbness on top of her head and in her hand. She has had improvement with her twitching since taking Taurine, an amino acid supplement. Her headaches have been debilitating at times. She has found a great deal of relief from taking Petadolex. She has an autoimmune episode after nearly every "bug" she catches.... these have included toxic synovitis, myositis, costochondritis, pilomatrixomas, and so on.

Number responding = 3 / percent responding = 2% of total respondents (143).

Page 102. VI. Any Other Affected Family Members cont.: Comments for facial and/or optical asymmetry

1. Please enter any comments for other family members with facial and/or optical asymmetry:

Responses (3)
Please see previous comments. Also, our great aunt also had a squint and so does my son.
My son is not diagnosed because a professor told me it would be a problem according to insurance. Our general practioner realizes that my son is affected - but it is up to me whether a diagnosis is doing anything good for him.
My grandmother thought it was normal, nobody ever noticed it except me. Saw in the mirror one day that her face looked liked mine. Small atrophying in lips and chin and brown spots all over neck. She thinks it may be from a small stroke that was not diagnosed.

Number responding = 3 / percent responding = 2% of total respondents (143).

Page 99. VI. Any Other Affected Family Members

2. Are there any other family members with neurological symptoms?

Symptoms	Responses (124)	% of Number responding
Yes	19	15%
No	105	85%

Number responding = 124 / percent responding = 87% of total respondents (143).

Note: If "Yes" was selected at the above question 99. VI. 2. "Are there any other family members with neurological symptoms?" then the following section (Page 103) would apply.

Page 103. VI. Any Other Affected Family Members cont.: Comments for neurological symptoms

1. Please enter any comments for other family members with neurological symptoms:

Comments (17)
Brother has special needs. He is a slow learner with poor short term memory.
Mom - very rare eye disease where pupil does not dilate correctly - only affects right eye (left eye "normal").
My baby sister has grand mal seizures.
My son has Tourette Syndrome. He's 11 years old and was diagnosed at age 7.
My son has Jacksonian Epilepsy and migraines.
Grandma - stroke Mother - thyroid problems
Dad had Parkinson's disease.
I have a sister with MS (Multiple Sclerosis).
"electric shocks"
Biological half brother with Autism and 2nd cousin with severe epilepsy (2 brain surgeries).
Aunt has MS (Multiple Sclerosis).
Father had motor neurone disease Grandmother had a brain tumor cousin (Joy) grandson has autism cousin (Jenny) son had Tourette Syndrome cousin (Corrie) has Graves' disease
Dad has brain tumour and cluster headaches (but these are unrelated to each other). Uncle had surgery for some type of face pain.
Depression, Bi-Polar
My father has an unusual atrophy of his eye muscles that control coordination of vision and eyelids. He sees double and does not have the muscle strength to keep eyelids open (they are essentially sewn open).
My mother has shingles on her face. I don't know if this qualifies as neurological, but I know some of her nerve treatments have required a neurologist.
Father epileptic Sister has Sjogren's Syndrome

Number responding = 17 / percent responding = 12% of total respondents (143).

Page 104. VII. Miscellaneous Questions (page 1 of 2)

1. Are there any non-Romberg family members who have an autoimmune disorder?

Autoimmune disorder	Responses (125)	% of Number responding
Yes	35	28%
No	90	72%

Number responding = 125 / percent responding = 87% of total respondents (143).

Note: If "Yes" was selected at the above question 104. VII. 1. "Are there any non-Romberg family members who have an autoimmune disorder?" then the following section (Page 105) would apply.

Page 105. VII. Miscellaneous Questions cont.: Others with an autoimmune disorder

1. Please select any Autoimmune disorders for non-Romberg family members:

Autoimmune disorders	Responses (96)	% of Number responding
Addison's disease (adrenal)	0	0%
Ankylosing spondylitis	0	0%
Autoimmune Thyroid disease	6	17%
Arthritis	14	39%
Celiac disease	4	11%
Chiari Malformation	0	0%
Chronic Fatigue Syndrome	3	8%
Crohn's disease	2	6%
Fibromyalgia	5	14%
Goodpasture's Syndrome (lungs, kidneys)	0	0%
Graves' disease (thyroid)	2	6%
Guillain-Barre Syndrome (nervous system)	1	3%
Hashimoto's thyroiditis	2	6%
Hughes Syndrome (antiphospholipid)	0	0%
Inflammatory bowel disease	2	6%

Continued on the next page.

Autoimmune disorders	Responses (96)	% of Number responding
Lupus (SLE)	3	8%
Mixed Corrective Tissue disease	0	0%
Multiple Sclerosis (MS)	4	11%
Polymyalgia Rheumatica (large muscle groups)	0	0%
Raynauds Phenomenon	6	17%
Scleroderma (skin, intestine, less commonly lung)	1	3%
Scleroderma, linear	0	0%
Scleroderma, localized	1	3%
Sjogren's Syndrome	1	3%
Systemic sclerosis	0	0%
Temporal Arteritis / Giant Cell Arteritis (arteries of the head and neck)	0	0%
Thyroid problems	11	31%
Type 1 Diabetes Mellitus	6	17%
Ulcerative colitis	3	8%
Vitiligo	6	17%
Other (Specify)	13	36%

Number responding = 36 / percent responding = 25% of total respondents (143).
Note: This was a multiple choice question.

Page 105. VII. Miscellaneous Questions cont.: Others with an autoimmune disorder
1. Please select any Autoimmune disorders for non-Romberg family members: Other (Specify)

% of Total Responses	Responses (13)	Other - Select Any Autoimmune Disorders
1%	1	Meniere's disease
1%	1	Psoriasis and Psoriatic arthritis
1%	1	severe allergies
1%	1	eczema
1%	1	thyroid gland
1%	1	Lupus Erythmatosis
1%	1	Psoriasis
1%	1	rheumatoid arthritis
1%	1	Granuloma Anulare
1%	1	Unsure, it was my grandad who is now deceased.
1%	1	cancer
1%	1	Neuropathy
1%	1	liver disease - autoimmune

Page 105. VII. Miscellaneous Questions cont.: Others with an autoimmune disorder
2. Comments for Autoimmune disorders:

Responses (10)
A few older adults in parent's families have been diagnosed with: 1. Raynauds (Mild) 2. Vitigilo 3. Psorasis 4. Arthritis
My mother has one form of Lupus, while my daughter has SLE (systemic lupus erythematosus). My baby sister has grand mal seizures and the doctors can't figure out why she has them.
Paternal grandfather has diabetes. Paternal uncle has Crohn's.
I myself have Raynauds Phenomenon.
Both my husband and I have several family members with autoimmune disorders.
Lichens Planus
1 illnes for three of my family members
paternal aunt
Myself and other children have problems.
My mother (my daughter's grandmother), and a neice on her father's side has ITP (idiopathic thrombocytopenic purpura).

Number responding = 10 / percent responding = 7% of total respondents (143).

Page 104. VII. Miscellaneous Questions (page 1 of 2)
2. Are there any other family members who have headaches or neurological symptoms limited to one side of the head?

Have Symptoms	Responses (124)	% of Number responding
Yes	13	🟥 10%
No	111	🟦 90%

Number responding = 124 / percent responding = 87% of total respondents (143).

Note: If "Yes" was selected at the above question 104. VII. 2. "Are there any other family members who have headaches or neurological symptoms limited to one side of the head?" then the following section (Page 106) would apply.

Page 106. VII. Miscellaneous Questions cont.: Others with neurological symptoms
1. Please comment on other family members with neurological symptoms limited to one side of the head:

Comments (10)
Mother is experiencing severe migraines with aura.
The left frontal lobe on my sister's head.
Dad had sinus headaches.
Mother has migraines usually affecting only one side of head.
My mom has a history of migraines.
Migraine
Ashton's maternal Grandmother's migraines were usually limited to one side of her head.
Cluster headaches- brother
Migraine headache
Cousin has migraines, pain located on right side of the brain, left side of the body goes numb for a while. I have had the same thing, it seems to be over now.

Number responding = 10 / percent responding = 7% of total respondents (143).

Page 104. VII. Miscellaneous Questions (page 1 of 2)

3. Has the affected person tried any diets (e.g. anti-inflammatory) to help with the Rombergs symptoms?

Any diets	Responses (123)	% of Number responding
Yes	11	9%
No	112	91%

Number responding = 123 / percent responding = 86% of total respondents (143).

Note: If "Yes" was selected at the above question 104. VII. 3. "Has the affected person tried any diets (e.g. anti-inflammatory) to help with the Rombergs symptoms?" then the following section (Page 107) would apply.

Page 107. VII. Miscellaneous Questions cont.: Tried any Diets (e.g. anti-inflammatory)

1. Please give a general description of this diet:

Comments (8)
Anti-inflammatory vegan diet
It was an injectable treatment at every 3 days. I had the injection for almost a year .... and pills... every day a pill.
I started eating semi-spicy foods because of a friend, and I noticed that if I had a 'dent' headache when I ate this, the pain would subside some, even if just for a short time, there was some distinct relief. I started slowly increasing the spiciness and found that for medicinal purposes to have the spice just enough to be tolerable but still feel the heat permeate through the skull and causing the body to heat up to a slight perspiration is ideal in relieving pain. I often go to the Malaysian restaurant for curry dishes and the Thai restaurant for Tom Kha soup. They have come to know me there as the woman who comes in not only for good food but for medicinal reasons.
Dairy free diet - (not so good at following as am a chef by trade) - and have no tolerance for pork, also watch that I don't eat too much prepackaged processed foods. (just because there are too many unknowns with processed foods, feel generally better eating what my grandparents ate.
Trying to follow an anti-inflammation diet... more vegetables, olive oil, omega 3, etc.
Low cholesterol diet
Weight control
N/a

Number responding = 8 / percent responding = 6% of total respondents (143).

Page 107. VII. Miscellaneous Questions cont.: Tried any Diets (e.g. anti-inflammatory)

2. Please comment on the results of this diet:

Comments (8)
Had fewer headaches, but was always very tired.
Nothing changed ... I did not feel better.
It works for me, at least to some degree. And when you're in pain, any amount of relief is a blessing.
When I stick to it, it works to reduce ulcerative colitis without taking drugs.
Nothing yet
My first (and only) episodes of TIAs occurred when my cholesterol was quite high. This was 5 years ago and since then I have followed the low cholesterol diet and have not to date had a recurrence.
Stress and lots of mood changes.
N/a

Number responding = 8 / percent responding = 6% of total respondents (143).

Page 108. VII. Miscellaneous Questions (page 2 of 2)

1. Has the affected person seen any improvements of the Rombergs symptoms with lifestyle changes?

Changes	Responses (119)	% of Number responding
Yes	21	18%
No	98	82%

Number responding = 119 / percent responding = 83% of total respondents (143).

Note: If "Yes" was selected at the above question 108. VII. 1. "Has the affected person seen any improvements of the Rombergs symptoms with lifestyle changes?" then the following section (Page 109) would apply.

Page 109. VII. Miscellaneous Questions cont.: Lifestyle Changes

1. What if any life style changes have been helpful:

Comments (21)
Do not over do things - get more rest. PRS symptoms seem to be exacerbated by being run down/tired.
Exercising and eating healthy foods
Acupuncture
Good rest and diet-try to keep stress low. Quit smoking!
Resting when I need to and not just plowing through the fatigue unless I have to. Taking a multi-vitamin everyday. Eating healthy foods.
Good diet, less stress
Cutting out sugar and hydrogenated corn syrup has reduced the discoloration.
Always helpful to reduce inflammation through foods and herbs, with cases of autoimmune situations.
Stay healthy and having a good attitude helps. If I'm not active for a while I feel pain sometimes.
Regular aerobic exercise
A better diet has helped overall.
For me it was more of being able to cover up my left side with make up and how to do my hair a certain way to cover up my forehead and not cause a lot of attention towards it.
I use to go to the gym regularly but found that I lost fat on my face more quickly on the affected side than the normal side. When I stopped, the fat on my normal side returned but not on the Rombergs affected side.
Leading healthy lifestyle as in more exercise, relaxation and less stress - I have found that when I can do this, I do not have any symptoms such as face tingling, body jumps but when I get stressed they come back again. I find that I grind my teeth at night and end up with a lot of pain the next morning in teeth and other areas of my face. But doing a relaxation session with progressive muscle relaxation prior to going to bed solves this problem and the effect is instant.
All things that keep negative stress away from me, means good partnership, good friends and family, good working conditions, being content with my life and so on. Healthy life-conditions (no smoking, no drugs, healthy food, etc.)
Continued on the next page.

Page 109. VII. 1. Comments (continued)
Exercise!
Yoga, healthy nutrition (no preservatives, no processed, no hormones, organic foods). Exercise and different aproaches to relieve anxiety and stress.
Cut out all high-fructose corn syrup and cutting down on processed sugar has reduced and faded the dark discoloration on his face.
Decrease stress Eat healthy Exercise regularly
Do not eat sugar.
Watching what she eats for migraines.

Number responding = 21 / percent responding = 15% of total respondents (143).

Page 108. VII. Miscellaneous Questions (page 2 of 2)

2. Have any health professionals that you have consulted suggested any theories of what they believe may cause Rombergs?

Any theories	Responses (125)	% of Number responding
Yes	20	16%
No	105	84%

Number responding = 125 / percent responding = 87% of total respondents (143).

Note: If "Yes" was selected at the above question 108. VII. 2. "Have any health professionals that you have consulted suggested any theories of what they believe may cause Rombergs?" then the following section (Page 110) would apply.

Page 110. VII. Miscellaneous Questions cont.: Believe May Cause Rombergs

1. Please comment on the health professional's theory for the cause of Rombergs:

Comments (20)
That it could have been triggered by trauma.
Some say it's caused from the enviroments, something I was exsposed to. Some say it's because I was a very sick kid and had a low immune system. Some say it was caused from the swimming accident that triggered it.
Lyme disease
1. Dr. Tsai: Multifactoral autosomal dominant trait, with the multi-factors possibly being predisposition of autoimmune disease, trauma (even very mild trauma), disease (virus or bacteria), stress (physical or emotional) and/or hormones (either naturally occurring hormonal changes or drug induced hormonal changes such as birth control pills) 2. Vascular constriction or degeneration
Trauma Genetically predisposed
The rheumatologist I see says it is an autoimmune disease. He does not know what the trigger is. I know some members believe it is neurological in origin but my doctor disagrees with that. He seems sure it is autoimmune.
An FNP (Family Nurse Practitioner) colleague said PRS is a gentic defect effecting the CNS (Central Nervous System).
Inflammation sets off the body's response (something threw off the balanced biochemistry of the system) and we need to reset it some way.
My surgeon felt it is caused by some sort of interruption of the flow of blood to the affected area.
Perhaps x-rays taken while my mother was pregnant with me during her 3rd trimester.
My doctor has a theory that it was a connection with rabbits, but overall, there is not much known about the disease.
Trauma to the side of the face/head not affected by Rombergs.
Same as mine, there is a relationship between the illness and the injury.

Continued on the next page.

Page 110. VII. 1. Comments (continued)
One doctor said that some people were just more prone to getting it. No real cause was given.
Possible autoimmune
Trauma to the face during childhood Pregnancy and childbirth Overlapping autoimmune diseases
I saw a speech pathologist a few years ago and she specializes in treating stroke patients with facial weakness and asymmetry. She had pioneered the use of a microfacial stimulation unit or special exercises in treatment of this condition and had been having success. She came up with a theory on the cause of Rombergs - It was all to do with the blood flow to the muscles. It is very complicated but at the time it made absolute sense to me but now I find it hard to put it into words so I have attached a document (previously circulated) with her theory and possible treatment. I did try the exercises but had such a weird reaction that I was wary of following through, although she thought my reaction was a positive sign. The fact that she was in a different State than me made it all very difficult. I tried to get doctors in my home State to follow through but had no luck - I would love for a doctor who is familiar with Rombergs to explore this theory and possible treatment.
Not really a cause but that it is due to the secondary nervous system - but not quite sure why it behaves as it does.
Mayo Clinic: Related to thyroid problems and/or diseases. Our family has underactive thyroid issues.
Putting a crown on a tooth on PRS side triggered it from that very day. That's what doctors believe.

Number responding = 20 / percent responding = 14% of total respondents (143).

Page 111. VIII. Closing Survey
1. Any closing comments:

Comments (63)
Thank you!
Thank you for all that you are doing to research this terrible disease.
I would love to get a copy of this survey once complete, to give to my doctors here in San Diego, CA. My husband is in the military and a lot of the military doctors are very curious about me and want to learn more.
Thank You to all who have helped devise this survey. RESEARCH is the route to answers.
I am the only member in my family who has Romberg's, but not the only one with an autoimmune disease.
Thank you so much for making this survey. I hope it gets to you on time!
Thank you for asking me to participate in this survey.
I am very disappointed by the medical system in my country. No one cares........ I was never informed about my disease. The drugs they gave me were small experiments ("let's see if u take that what happens!") and so on....I thank this site because here I found the support I need and information.
I am still at a loss as to what could have caused it, and why it is affecting me at 26 years old. I am having a lot of anxiety now about becoming pregnant (which I was hoping to be in the next year). Many people have expressed concern about changes in their pregnancy.
Thank you for taking the time to put this all together.
This was very user friendly. You did a great job creating this survey. Thank you for doing this. I hope it will be helpful to many.
Gerri - sorry this is late - have just had second fat injection operation. Finding this group has been a godsend for me - I had always thought I was the only one of my kind.
I only had surgery on 10-02-09. It may be too soon to tell whether the fat will remain. It looks better and I will be going for a check-up in 6 weeks time. Will keep all posted.
Thanks for doing this Gerri!!! It must have taken a lot of your time. I do hope something common to all of us Rombergers will reveal itself! Thanks again to you & Marilyn. Love Jane (UK)!
Hopefully, someday we will find the cause of this syndrome. Thank you for the survey/study.
Thank you for taking the time and effort to create this survey as well as the website. Like so many of our members, I felt devastated and lost when my son was diagnosed with PRS and did not know where to turn. Initially the medical community told me there was nothing we could do except to wait for this disease to "burn itself out" and then at that time proceed with surgery. This "Connection" is the lifeline to a body of patients' experiences and knowledge of this dreadful, heartbreaking "syndrome". I hope one day the "Romberg Connection" will be put out of business because someone will have discovered the cause and then the cure for this disease.
This disease has so much more to it than this survey dwells on, like the everyday struggles we all go through with doctors and the always feeling bad and bloom (fatigue feelings) we have all day and how that effects our everyday life and how others look at us... what research we have done as a person... What ya'll have done is great but it is a shame that doctors aren't doing this or helping at all. This is crazy how the medical department treats people like us. Millions go un-diagnosed a year and millions like us don't get any help at all. Nichole
This disease has so much more to it than this survey dwells on, like the everyday struggles we all go through with doctors and the always feeling bad and bloom (fatigue feelings) we have all day and how that effects our everyday life and how others look at us... what research we have done as a person... What ya'll have done is great but it is a shame that doctors aren't doing this or helping at all. This is crazy how the medical department treats people like us. Millions go un-diagnosed a year and millions like us don't get any help at all. Nichole

Continued on the next page.

Page 111. VIII. 1. Comments (continued)
My GP (General Practitioner) says I am a mutant.
We need to become politically active to promote research for orphan diseases'. With the change in administration (at last) in DC, now might be time we have leaders who will listen.
My thought is that PRS, Localized Linear Scleroderma, Trigeminal Neuralgia, Silent Sinus Syndrome, and Morphea, along with various unilateral eye, ear, nose and mouth symptoms are all part of a one-in-the-same atrophic process, and that they are given a diagnosis based upon which medical specialty first makes that diagnosis. For example, a rheumatologist is going to name it differently than a neurologist will. I liken the atrophic process to a large snow storm. It's all the same weather front, but it affects different areas in different ways, and to varying degrees. Some are affected by hair loss, some by vision or eyeball changes, some by neurological symptoms, others by dental or dermatological changes, etc. but it is still all the same "storm". My personal feeling is that we should not limit our research or thinking just to PRS, but we should include these other atrophic cranial/facial diseases in our research because they may all be the same beast, just named differently, depending on who labeled it. If we had been diagnosed with Localized Scleroderma, that might be all we were looking into and we would know nothing of PRS, so we should branch out a bit in our research.
Thanks!
I am a member of Marilyn's Yahoo group, but have been silent for a few years now. The reason being that the Romberg's has progressed and I am very depressed because of it. I should be looking to the group for support during this time, but do not want to discourage anyone who feels that theirs will not come back again. So I read the posts from the group but do not participate for this reason. This is such a devastating disease and nobody can know how it feels or what we have to go through every day. I am amazed that some people in the group post their photos. I stay away from photos all my life because of this. I had my surgery at NYU (New York University) and wish I could get the strength to get it done again. I appreciate you taking the time to study this disease, I feel there are a lot more people who have it and do not know or their doctors do not know what to call it. I actually know more than most of my doctors about Rombergs and all of the symptoms. I hope and pray that someday they will find a way to stop it and find out what causes it. Thank You again for doing this study.
My Dad had Parkinson's disease - not sure if any link. Thanks for doing the survey - look forward to seeing the results!
Thank you Gerri & Marilyn for all that you do with the Connection! Brian
SUCCESS with the survey
This affliction has not affected my life like some and because no one took it seriously my GP (General Practitioner) only found out in the last couple of weeks (and didn't really have anything to add), have not really taken it too seriously myself. Maybe I should have or would have if more professionals took an interest instead of "Oh I don't know... maybe it will go away, I don't really know". I just know I have one switched on dentist who recently passed away from breast cancer and I will forever be grateful for her interest. If anything I add to this helps you guys find out how this works and what can be done to stop the disfigurement and self esteem issues and the very real physical ailments that this thing causes, that would be great. Just because someone doesn't know what something is, doesn't mean it will just go away. Good luck I hope your survey yields something that parents of new PRS people can hold onto.
Great job on the survey. Thanks for all of your hard work. Much appreciated!
Our daughter is adopted (at 5 weeks old) so we really do not know her family history. So the questions regarding family I answered no to but they are subject to the unknown.
Great job Gerri & Marilyn. Thank you for doing so much to help PR.
Thank you for the great effort in putting this together!!!
Good luck & thanks.
I just joined the Group after just being diagnosed. I'm 51. My answers are somewhat incomplete because I don't have all the data. For instance I have many dental problems. Now...I'm making the connection that these problems could be from the Rombergs. I have always had trouble sleeping. So is this part of the Rombergs? So I'm in the process of back tracking my medical history as I learn more about the symptoms. I have only seen the reconstructive surgeon once and that was when I was diagnosed.
Continued on the next page.

Page 111. VIII. 1. Comments (continued)
Thanks for organizing the survey…
Thank-you for your dedication and hard work toward this effort.
Deciding to start my daughter on meds was a huge choice to make but I am so glad I did, vast improvement and activity has stopped. No side effects and my daughter is so pleased with the results. A very positive choice.
Thank you for doing this. I am extremely eager to see the results. Please let me know if you need additional information and/or if you see any pattern to the treatments that are working. There has to be an answer and a cure.
*Educate yourself and others about Parry Romberg Syndrome in a way that is comfortable for you. There is less fear or curiosity when the facial "difference" has a name. *Have a print-out that explains the disease and bring it to doctor visits. See if you can fax or email it to them before the appointment, so they have a "heads up". Remember that most doctors are still unfamiliar with this disease. If they are not willing to learn about it...find another doctor. *Educate yourself about the disability laws, especially if you have a child with PRS in school. Section 504 is an anti-discrimination, civil rights statute that requires the needs of students with disabilities to be met as adequately as the needs of the non-disabled are met. If your child has headaches, vision problems, attendance issues, they may be helped with a 504 Plan. Be an advocate for your child. Tell your school you would like to set up a meeting to talk about putting your child on a 504 Plan and making appropriate accommodation or modifications that can help your child to be successful. Just Google: Section 504 to learn more. *If at all possible, occasionally meet with families or individuals who share the same rare disease... for friendship and support. These are the people who truly GET IT. *If you are a parent of a child with PRS, talk about it with them. From the experiences we heard, one of the most painful things a parent can do is IGNORE OR DENY that there is a problem. *Support the Parry Romberg organizations that are out there now. *Above all...SUPPORT EACH OTHER! NO ONE HAS TO FEEL ALONE WITH THIS DISEASE."
Thanks for putting the survey together! It will be interesting to see the results.
Somewhere along the line I must have missed the question about TIA's (transient ischemic attacks) - I did not tick the "brain" thinking it meant mental state so I missed any follow through on that. I have suffered from TIA's or possibly a blood clot behind the eye - I take Cartia (low dose aspirin) every second day plus fish oil. Marilyn and Gerri - thank you so much for putting this together - we just need a doctor or a researcher to use this information to find a cause and more importantly treatment and a cure.
Am sorry I am not able to provide more telling info. We go around and around wondering what the triggers are/were. Unfortunately, we (me and child) have a situation that may be the "perfect storm" as we had a polluted area, atypical pneumonia and tick bites in a short window of time. Doctors say I am too anxious, but don't know how not to be.
I tried to answer as good as I could (German origin). So the only treatment is 20 years ago, I still cannot remember very well. If there are any problems in understanding my comments, please feel free to contact me.
Thank you for creating this survey! I truly hope we can all help to understand Rombergs better.
Survey was easy to use. You did a great job putting it together.
Some questions could be a little clearer.
I just wish that there was some way to diagnose this so that those of us who are parents don't have to keep looking at our young children's faces every day for signs of PRS to see if they've appeared, or if they are going to appear. Of course, that's probably just me. I am thankful that I got it when I did in my life, because I'm not sure that I would have been able to.
We have been to doctors who have recommended Methotrexate and pulse IV steroid treatment. At this time, we are looking for alternatives. We are flying to NY to see Dr. Siebert about surgery. Hoping for surgery this summer!
Continued on the next page.

Page 111. VIII. 1. Comments (continued)
Good luck with your survey.
Very interesting survey. My foot is a particular issue for me as the tissue loss means I can no longer run or walk a long distance. This seems unusual from other Romberg’s sufferers I have spoken to, so am very interested if any others have problems with their foot.
Marilyn and Gerri I can't thank you enough for your hard work and support.....and I know you put in many, many hours into creating this thorough survey. I am very impressed with the survey. I believe that the information from this survey will be helpful to scientific/medical professionals who work to investigate Rombergs and search for clues to prevent/control/conquer the condition. I would be happy to provide any additional information.....full name, address, etc..... if ever requested. There is hope in sharing information. I do feel that my Romberg's may have been triggered by dental work. After a recent conversation with my primary care physician, I am going to schedule an appointment with an "Infectious Disease" physician. My doctor does not know that I will learn anything that might be helpful to me, but he does agree with my feeling that since I have lichen planus, an Autoimmune disorder, which affects my whole mouth, and Parry Romberg, which affects my face and tongue, it is a reasonable idea to follow up with someone who may deal with Autoimmune problems. I will share with you anything of interest that I learn. The Romberg group, the e-mails, the contacts, mean a great deal to me. I do not feel alone.....and that is very comforting. It is through the group that I found a wonderful plastic surgeon....Dr Linton Whitaker of the University of Pennsylvania. Thank you both very much. Beverly Pennsylvania
Needed to be wee little clearer with the questions. Don’t get me wrong they were good questions and covered everything but I actually had to reread several cause I didn’t readily understand them.
Thank you. I hope this helps. I filled this out in lieu of my daughter, so it is third-person limited.
Any and all help is appreciated for this dreadful condition. More research needed.
Great initiative to have this survey! This one was clearly about medical/technical issues; maybe another one on personal issues is a good idea? I think people suffer more from mental than from physical problems due to Romberg. Anyway, well done!!!
Thank you for continuing your commitment to all of us.
Thank you for what you're doing!
Hope some of this helps.
Thank you
I didn't answer some questions because they didn't apply to me. I hope this survey helps future treatment of this terrible disorder. It has been a very difficult and expensive thing to deal with.
I feel my symptoms are probably reasonably mild compared to a lot of people. At the onset of my symptoms, the specialist diagnosed it as scleroderma after a biopsy was done on my right thigh at the age of 8. It wasn't until I was in my late twenties that my local GP (General Practitioner) diagnosed it as Parry Romberg. He told me it was an unknown quantity but as my condition hadn't worsened from about the age of 13, he thought it had certainly stabilized. I have had a lot of back problems due to severe scoliosis and have had 5 back operations. The last one 12 months ago was the big one. Fused from L5-S1 to T9 with about 30 screws, rods on either side of the spine and 3" screws into the pelvis to stabilize the spine. Part of this could be caused by the atrophy in my back but my back specialists have never heard of this syndrome so we still wonder whether there is an association at all. If you need any more information please email me. Pip

Continued on the next page.

Page 111. VIII. 1. Comments (continued)
My daughter had lead poisoning as a toddler and I've always wondered if the Rombergs could be related.
We should translate this survey in Spanish and other languages for more participation. I for one would be more than willing to help.
Please let's help each other through this and hopefully with all the research and this survey, just maybe we can find hope and a cure.
I really did not thoroughly answer the questions about the meds and the surgeries. That's because I was completely discouraged by the inability of the Docs to try any alternative remedies & diets despite the fact that nothing they were doing was helping. Then they lobotomized her at Albany Medical and that made her brain damaged with right sided weakness but instead of trying to help, they discharged her ASAP because they didn't want to deal with what they did. She went down hill after that. I truly hope no other PRS has to ever go through the trauma that we did. Katie died May 27, 2008 at the young age of 39 from complications of PRS. Truly no one knew what to do for her.

Number responding = 63 / percent responding = 44% of total respondents (143).

Throughout this survey, when we use the term "Rombergs" or "Romberg" we are referring to: "Parry Romberg", "Parry Rombergs", "Parry Romberg Syndrome" and "PRS".

This is an unscientific survey designed to gather information from those who must deal with Rombergs on a daily basis.

For more information about The Romberg's Connection
please email us at:
rombergs@hotmail.com

Last Updated May 5, 2009