# AN IN-DEPTH LOOK AT C.R.P.S – FROM DIAGNOSIS TO TREATMENT

PHILIP GETSON, D.O.
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# DIFFERENTIAL DIAGNOSIS

- Diabetic and small-fiber peripheral neuropathies
- Entrapment neuropathies
- Thoracic outlet syndrome
- Discogenic disease

- Deep vein thrombosis
- Cellulitis
- Vascular insufficiency
- Lymphedema
- Costochondritis
- Brachial Plexopathies

Raja SN et al. *Anesthesiology.* 2002;96:1254-1260.

#### THE DISEASE

The pain is profound, chronic and widespread. It can migrate to all parts of the body and vary in intensity. The pain has been described as stabbing and shooting pain and deep muscular aching, throbbing, and twitching. Neurological complaints such as numbness, tingling, and burning are often present and add to the discomfort of the patient. The severity of the pain and stiffness is often worse in the morning. Aggravating factors that affect pain include cold/humid weather, non-restorative sleep, physical and mental fatigue, excessive physical activity, physical inactivity, anxiety and stress.

Additional symptoms may include: irritable bowel and bladder, headaches and migraines, restless legs syndrome (periodic limb movement disorder), impaired memory and concentration, skin sensitivities and rashes, dry eyes and mouth, anxiety, depression, ringing in the ears, dizziness, vision problems, Reynaud's Syndrome, neurological symptoms, and impaired coordination, sleep disturbance and fatigue

The preceding two slides are directly from the

#### National Fibromyalgia Website

## In MY opinion Fibromyalgia <u>IS</u> CRPS

#### REFERENCES

### Is Fibromyalgia a Generalized Reflex Sympathetic Dystrophy?-

M.Martinez-Lavin- "Clinical & Experimental Rheumatology", 2001: 19:1-3

# Fibromyalgia and the Complex Regional Pain Syndrome: Similarities in Pathophysiology and Treatment

Richard J Wurtman – "Metabolism Clinical and Experimental"-59-2010 -837-840

#### **MULTIPLE SCLEROSIS**

I have studied nine patients, all female, who have been diagnosed with BOTH MS and CRPS.

Treatment was initiated to the MS by neurologists and ALL nine showed improvement of the MS symptoms while seven showed improvement of the CRPS

#### **SYMPTOMS**

- Sensory
   Burning pain
   Skin, sensitivity to touch
   Skin, sensitivity to cold or heat
- Vasomotor
   Abnormal skin color changes
   Abnormal skin temperature changes

#### **SYMPTOMS**

- Sudomotor / Edema
  - Abnormal sweating
  - Abnormal swelling
- Motor / Trophic
  - Limited movement
  - Weakness
  - Tremor
  - Dystonia
  - Neglect
  - Abnormal hair and/or nail growth

#### **Vasomotor Changes**



#### **Abnormal Sweating**



#### MOTOR DISTURBANCE-DYSTONIA



#### **NEUROGENIC EDEMA**



#### **ERYTHEMA**



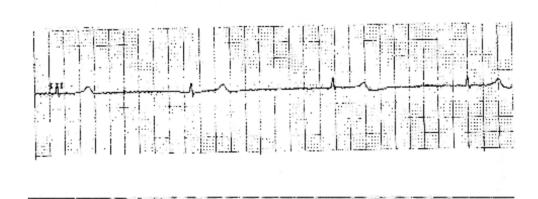


#### **EDEMA & ERYTHEMA**



#### ORGAN INVOLVEMENT

#### **CARDIAC**



#### CASE PRESENTATION

The patient whose EKG strip appeared on the previous slide was a 45 y/o female who presented in the ER following a syncopal attack. She had a history of Type II diabetes but blood sugar was 160. She had a history of thyroid dysfunction but TFS were all WNL. She had no other medical history save for her CRPS

### ARRHYTHMIAS

BRADYARRHYTHMIAS – Usually in female patients over 40

TACHYCARDIAS – Usually in female patient in their 20's

ATYPICAL RHYTHMS

#### **CHEST PAIN**

Many patients complain of chest pain that, following a comprehensive cardiac work-up proves to be a result of CRPS. The etiology appears to be sensitization of the Intercostobrachial nerve in patients with a brachial plexus injury

Atypical Chest Pain: Evidence of Intercostobrachial Nerve Sensitization in Complex Regional Pain Syndrome – Rasmussen et al – *Pain Physician 2009*; 12:E329-E334

A 50 year old male has a massive M.I. necessitating quadruple bypass surgery. The operation goes flawlessly and he awakens in more pain than when the surgery began — that pain accelerates and does not subside 12 years later.

#### VISUAL DISTURBANCE

DOUBLE VISION
BLURRED VISION
OCCULAR MIGRAINES
VISION LOSS
PHOTOPHOBIA
BURNING OF THE EYES

#### **HEADACHES**

# MIGRAINES(?) TENSION HEADACHES GREATER OCCIPITAL NEURALGIA



### • • • OTORHINOLARYNGOLOGIC

Patients report significant otophobia.

Recently there has been an increase in individuals reporting significant discomfort from <u>vibration</u>, specifically the bass of stereos even through walls and from adjacent motor vehicles while travelling.

- I have one patient who in addition to pain etc. began to stutter uncontrollably as the CRPS worsened.
- After on week of IV Ketamine he stopped stuttering altogether

#### DENTAL

Unfortunately dental disease is rampant in patients with CRPS

Part of this stems from dietary indiscretions, part from immune system compromise and part from the disruption of the dental nerve roots.

Perhaps the greatest reason is that the side effects of common medications prescribed for chronic pain lead to a change in lifestyle, poor oral hygiene, poor nutrition and a loss of saliva (dry mouth) that result in decay, periodontal disease and ultimately tooth loss

#### **DERMATOLOGIC**

• The most common finding apart from dry skin or hyperhydrosis is <u>neurodermatitis</u>. This can occur randomly on any area of the body. Lesions have the appearance of small acne-type eruptions that itch for hours to days and disappear spontaneously. There is no specific etiology apart from the CRPS and no treatment save for topical low potency steroids or anti-histamines to reduce the itch. If scratched they will scar



#### **DERCUM'S DISEASE**

**Dercum's disease**, is a rare condition characterized by multiple, <u>painful</u> lipomas. These lipomas mainly occur on the trunk, the upper arms and upper legs. The understanding of the cause and mechanism of Dercum's disease remains unknown. Possible etiologies include: nervous system dysfunction, mechanical pressure on nerves, adipose tissue dysfunction and trauma.



#### LIVIDO RETICULARIS





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#### Gastrointestinal

Apart from the obvious acid peptic and irritable bowel symptoms, we have to deal with intractable nausea and vomiting. Endoscopically there may be some mild gastric irritation but generally the findings are minimal. Conventional treatment is rarely effective. The etiology is clearly **gastroparesis** and objectively identified via gastric emptying studies.

We have had great success with the endoscopic administration of Botox into the pyloric sphincter. In many instances one to three such injections have stopped the vomiting for prolonged periods of time.

- Our only two failures with botox were both females with intractable pain, nausea and vomiting and severe malnutrition and weight loss.
- They both ultimately underwent fundal plication surgery which was successful in reversing the GI abnormality
- THEY ARE SISTERS!

Another interesting finding is a number of patients with clinical and laboratory confirmed pancreatitis with no other etiology evident save for their CRPS

#### Other symptoms include:

Dysphagia, indigestion

Diarrhea / constipation (opioid induced)

Biliary dyskinesia

#### **Urinary**

Commonly, patients experience urinary incontinence, dysuria or inability/difficulty voiding. The condition is usually misdiagnosed as Interstitial Cystitis. The problem has responded marginally to conventional medications. Bladder pacemakers have been somewhat useful. Again, Botox injections into the pelvic floor have helped a great number of sufferers. Additionally, I have found that lumber epidural infusions of bupivicaine over a 5 day period works very well. Ketamine has resolved this to a small degree as well.

### Gynecologic

Menstrual dysfunction of all types is noted:

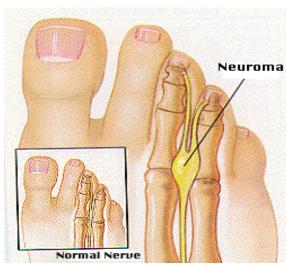
Polymenorrhea
Dysmenorrhea
Menometorrhagia
Secondary amenorrhea

### **OBSTETRIC**

An interesting finding is that patients in the third trimester of pregnancy (and some earlier in their pregnancy) become dramatically less symptomatic and many become <u>asymptomatic</u>. This lasts into and after childbirth and seems to be further extended by breast feeding. I currently have data on nine such cases and am exploring the hormonal shift that may be common to all of these individuals

### **PODIATRIC**

Morton's neuroma is a mechanically induced degenerative neuropathy predominantly affecting the second and third common digital nerves. It is not actually a tumor but a thickening of the tissue that surrounds the nerves leading to the toes. It is eight to ten times more likely to occur in women than in men and most prevalent in middle aged women



### **VERTIGO**

Vertigo is common as an early symptom Sometimes it is positional but mainly it is movement related

Treatment is based upon reducing the CRPS symptoms and occasionally meclizine helps make the vertigo tolerable pending the improvement of the disease process

#### **SYNCOPE**

"Syncope is common in patients with CRPS especially with lower limb involvement. Autonomic dysregulation of the lower extremities leads to sympathetic vasoconstriction and venous pooling which can predispose these patients to syncope."

Syncope in Complex Regional Pain Syndrome – Smith et al. – *Clinical Cardiology* 34.4; 222-225 (2011)

### Syncope-Case presentation

A 44 year old female with long standing history of CRPS is involved in a MVA which accelerates her symptoms. She further injures her brachial plexus in the accident and has classic symptoms of that sub-division of CRPS. However she begins to have "drop attacks" with increasing regularity.

Comprehensive work up with brain MRI, EEG, laboratory testing and carotid ultrasound all prove negative.

The solution proved to be immobilization in a soft cervical collar. Here is how that transpired......

### • • • COGNITIVE DYSFUNCTION

There have been an increasing number of CRPS patients with cognitive issues. Mostly these are STML, word retrieval & difficulty with expression. It has been theorized that this is medication related but it occurs in individuals who take virtually no meds. Current thoughts abound with no single answer surfacing as being definitive.

### CONSTITUTIONAL

- Lethargy
- Fatigue
- Weakness
- Sleep disturbance

#### **OTHER SYMPTOMS**

- Shortness of breath
- Inability to take a deep breath
- Neurogenic edema
- Muscle weakness/atrophy
- Endocrine dysfunction adrenal, thyroid, hormonal imbalance
- Gardner Diamond Syndrome –
   spontaneous bruising in uninjured areas

"Systemic Complications of Complex Regional Pain Syndrome"-Robert J. Schwartzman -Neuroscience & Medicine, 2012,3,225-242

### • • • ANXIETY & DEPRESSION

- Chronic pain = Depression
- Depression begets anxiety
- Benzodiazepines worsen depression
- SSRI's do not work
- SNRI's works marginally
- Currently my belief lies in the balancing of neurotransmittors

### **GENETICS**

A 37 year old female casino worker is struck by a "money cart" in the left lateral thigh and subsequently develops CRPS in that limb. It later migrates to the left arm.

One year later, her sister, a 35 year old police officer was broadsided in her patrol car while driving. The door handle impacts her left lateral thigh and SHE develops CRPS in the left leg which within months migrates to the left arm!

Currently I treat 18 families with more than one member who has CRPS.

There is one article in the literature that has studied genetics and CRPS – 31 families were studied, two families had five afflicted members, four families had four, eight families had three and 17 had two.

Familial occurrence of complex regional pain syndrome – deRooiji et al – Eur j Pain – 2009 – Feb; 13(2): 171-177

### THERMOGRAPHY

### "HEAT PICTURE"

# Autonomic Disturbed Function In The Case Of CRPS

A hallmark of CRPS is an excessive vasoconstriction of blood vessels that can cause cold hands and feet.

Because CRPS produces these neurovascular changes, infrared imaging, in a number of studies, has demonstrated a high degree of accuracy picking up these symptoms.

 Validation of Thermography in the Diagnosis of Reflex Sympathetic Dystrophy

Bruehl, et al. The Clinical Journal of Pain Vol. 12 (4)

Dec. 1996, pp. 316-325

 Long term skin temperature measurements – A practical diagnostic tool in complex regional pain syndrome

Krumova et al – Pain 140 (2008) 8-22

## What Causes (Cold) Emissions? (Hypothermic)

Vasoconstriction

**Tendons** 

Less Muscle Mass

Nerve Atrophy & Nerve Dysfunction

Sympathetic Stimulation

**Increased Fibrous Tissue** 

**Chronic Injuries** 

**Chronic Spasm** 

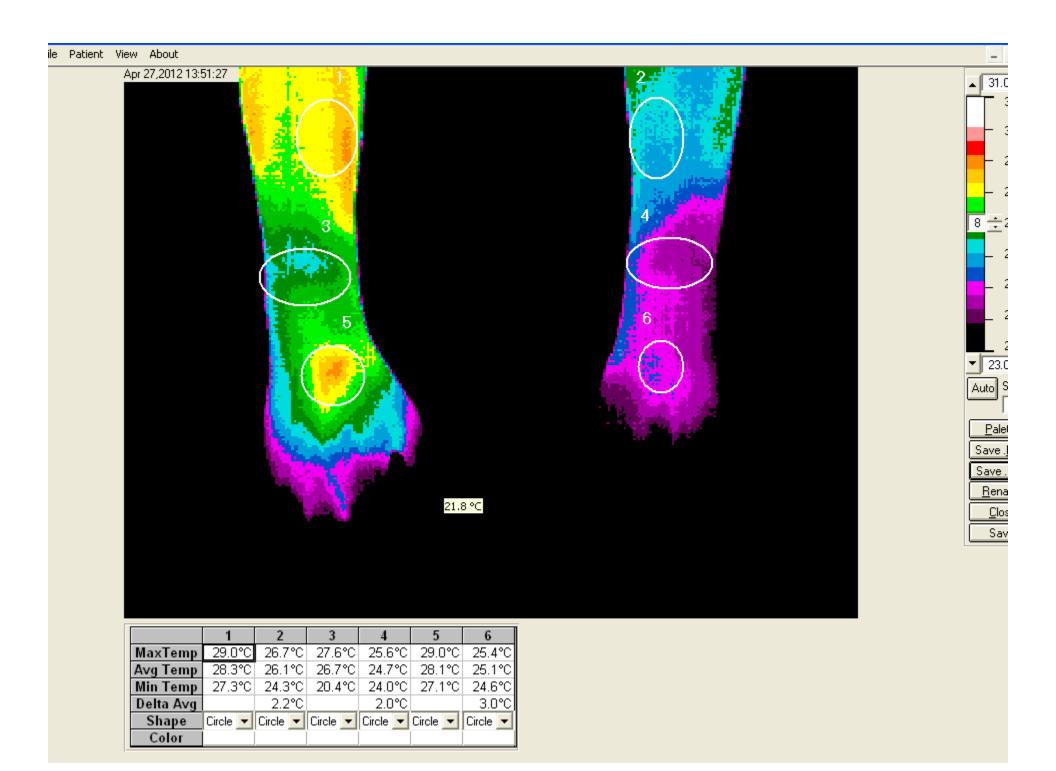
Disuse & Atrophy

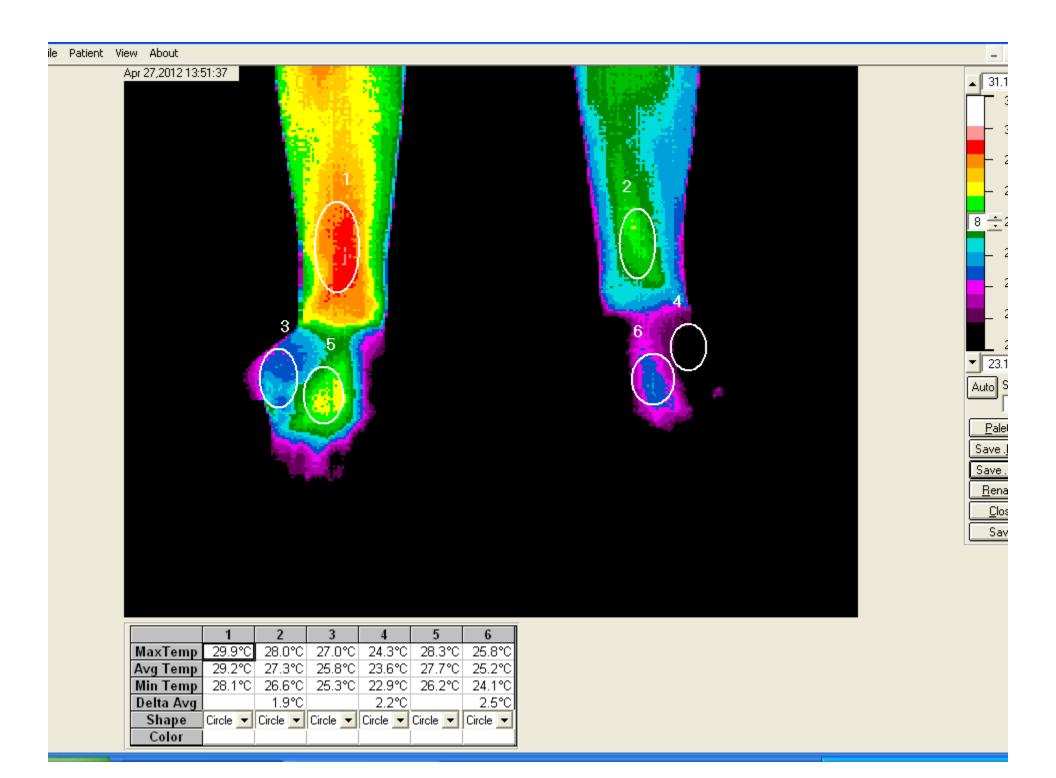
Muscle Inactivity

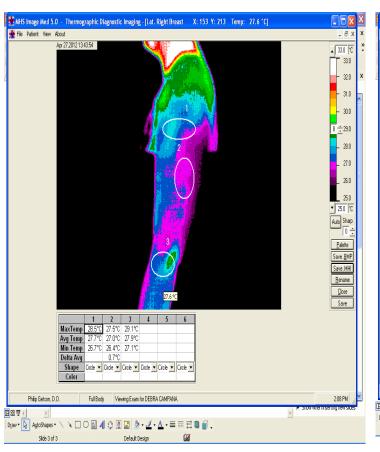
**Chronic Scarring** 

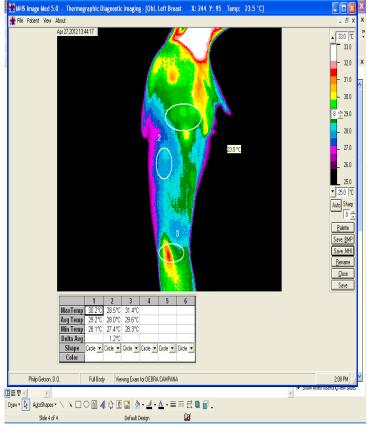
#### **CASE HISTORY**

A 40 y/o female was seated on the third row of a football stadium at a charity event when one of the players kicked a ball into the crowd. A fan threw the ball down attempting to reach the field but instead impacted the patient who, in an attempt to prevent getting hit in the face put her left arm up and was struck by the tip of the ball, fracturing her hand.









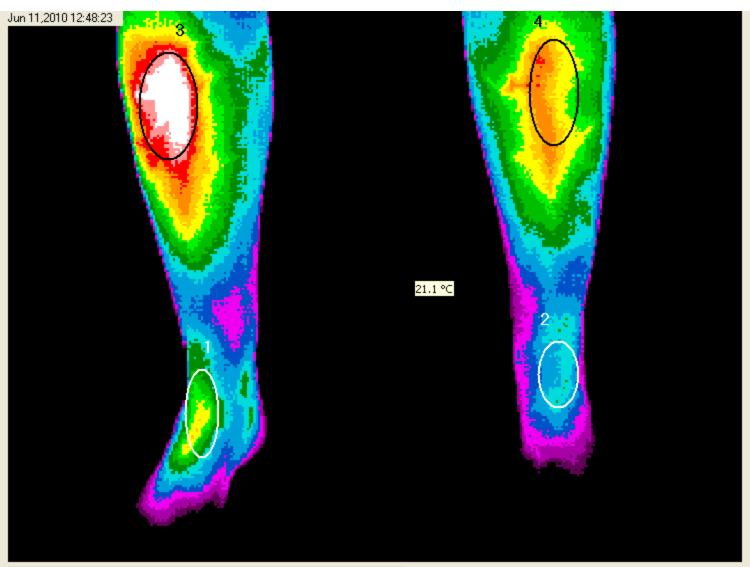
It has long been my opinion that thermography has a "predictive" value in the evaluation of patients with CRPS

The following is one example

### CASE HISTORY

A 43 year old female was an airline passenger wearing sandals. In the landing process she cut her *foot* on a piece of wire or metal that was sticking up from the plane

When examined in my office 2 ½ years later she complained of foot and ankle pain with a "squishy feeling."

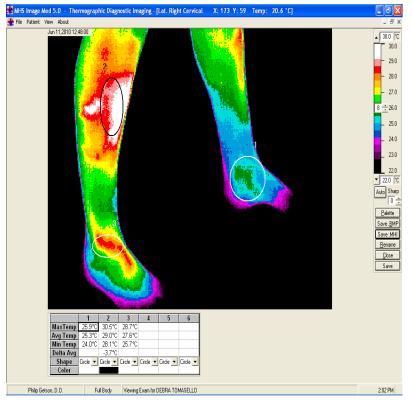


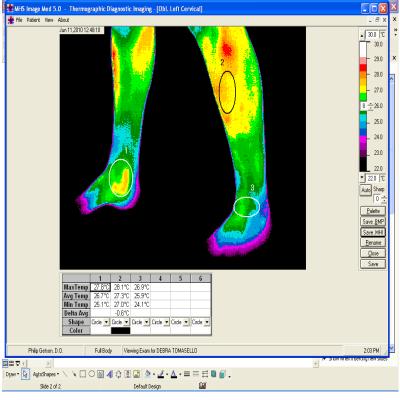
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Avg Temp	27.2°C	26.1°C	30.0°C	28.4°C		
Min Temp	23.4°C	25.1°C	28.5°C	27.3°C		
Delta Avg		1.1°C		1.6°C		
Shape	Circle 💌	Circle 🔻				
Color						

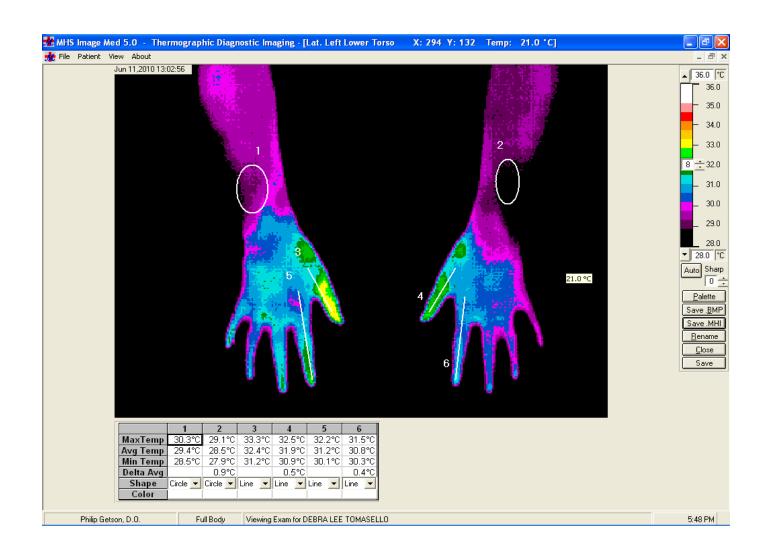
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### Each fact is suggestive in itself. Together they have a cumulative force.

**Sherlock Holmes** 

