

EFFICACY OF CERVICOTHORACIC SYMPATHECTOMY VERSUS CONSERVATIVE MANAGEMENT IN PATIENTS SUFFERING FROM INCAPACITATING RAYNAUD'S SYNDROME AFTER FROST BITE.

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Background: Raynaud's syndrome is a known complication of cold injuries. Stress, smoking and metabolic diseases may further aggravate the disease course. The purpose of this study was to determine the efficacy of Cervico-thoracic sympathectomy as compared to conservative management in severe Raynaud's syndrome after frostbite. **Methods:** This non-randomized controlled trial was conducted at Railway Hospital, Rawalpindi and Islamic International Medical Complex, Islamabad between January 1999 and June 2006. All patients sustained severe cold trauma in the mountain ridges of Himalayas in Kashmir. In all cases, an informed consent was obtained from patients and families. All operations performed were free of charges. Out of the total 48 patients who developed incapacitating Raynaud's syndrome of the upper limbs after frost bite, 17 patients underwent thoracic sympathectomy through anterior supraclavicular route. Remaining 31 patients were treated conservatively and were placed in the control group. Data was collected on pre-designed proforma and assessed using SPSS (version 11). Chi-square test was applied to assess the effectiveness of the two treatment modalities. **Results:** All operated cases initially showed improvement in symptoms and incapacitation. Among sympathectomised patients, 11 patients became symptom free and 3 patients showed mild but improved symptoms. Two patients after initial transient improvement developed incapacitating symptoms requiring further treatment, one patient developed gangrene of distal phalanx nine month after sympathectomy requiring amputation of the finger. Frequency of attacks and duration between the attacks reduced in all operated patients of cervical sympathectomy ($p < 0.05$) as compared to conservative management. **Conclusion:** cervical sympathectomy is a very effective modality of treatment in patients having severe Raynaud's disease of upper limbs secondary to frost bite.

Key words: Cervical sympathectomy, Raynaud's disease, frequency and duration of attacks, symptomatic relief, complications.

INTRODUCTION

Raynaud's syndrome is a vasospastic disorder affecting mainly upper limb vessels characterized by a sequence of events. The skin of the peripheral part of the limb (usually the fingers) first turns white, then blue and feels cold and numb. The affected skin may look slightly swollen. As the circulation improves, the affected areas may turn red, throb, tingle or swell. In a particular case all or some of these changes may occur. Raynaud's attacks may last from several minutes to several hours and may lead to complications like ulceration or gangrene of digits. Women are nine times more affected than men and it is more common in people who live in colder climates. Other factors like smoking and stress other than an exposure to cold can precipitate the attacks.¹⁻³

Signs and symptoms of Raynaud's depend on the frequency, duration and severity of the blood vessel spasms that underlie the disorder. Raynaud's can also affect other areas of the body such as nose, cheeks, ears and even tongue, but most frequently affects fingers and toes.^{4,5}

Primary Raynaud's syndrome occurs without an underlying disease or associated medical problem. In the primary form superficial necrosis is

very uncommon. Secondary Raynaud's syndrome (also called Raynaud's disease) usually affects both hands and feet. It is caused by an underlying problem and is less common than the primary form. Connective tissue diseases like lupus or Sjögren's syndrome, or conditions such as diabetes, thyroid disease, or peripheral vascular disease may be the underlying cause. Treatment of Raynaud's disease depends on its severity and the presence or absence of associated conditions. Medical treatment is often prescribed along with preventive measures to all patients with moderate to severe symptoms. There is no cure of Raynaud's syndrome or disease but symptoms can be modulated through medical means and in desperate situation by surgical means.⁵⁻⁷

Cervicothoracic sympathectomy has been described as a surgical option to minimize the symptoms and severity of this disease. The long term efficacy of the procedure is not established. However, failure of the long-term efficacy of the procedure does not prevent the surgeons to opt this mode of treatment. Most of the reported series mainly study the female population suffering from Raynaud's syndrome whether treated by medical or surgical

means. Moreover literature mainly stresses either on Raynaud's disease or Raynaud's syndrome caused by connective tissues disorders, thoracic outlet syndrome, occupational disorders and drug induced Raynaud's syndrome.⁸⁻¹⁰

This study was designed to determine the efficacy of open surgical cervicothoracic sympathectomy as a last resort of management in the patients with Raynaud's syndrome due to frostbite. Most of our patients were male. We also compared the long term efficacy of conservative management with cervical sympathectomy.^{10,11}

MATERIAL AND METHODS

In this study, a non-randomized controlled trial, total number of 48 patients suffering from Raynaud's syndrome were managed over seven and a half years in the hospitals of Islamic International Medical College Rawalpindi/Islamabad. All these patients sustained severe cold trauma in the Himalayas in the Northern Pakistan. All those patients affected by moderate to severe syndrome were offered cervical sympathectomy. Out of total 48 patients, 31 were managed conservatively by local application of 0.5% glycerol trinitrate paste and oral nifedepine 10–20 mg per day. Seventeen selected patients with severe Raynaud's syndrome were subjected to cervicothoracic sympathectomy through anterior supraclavicular approach.

In all cases, an informed consent was obtained from patients and families. All operations performed were free of charges.

All patients were routinely investigated by CBC, Urinalysis, Serum Urea, Glucose, and electrolytes and underwent thorough Doppler studies of the affected limb vessels, X-Rays of the thoracic inlet and clotting profile. Thirty-one patients were treated conservatively by proper counselling, local application of 0.5% GTN ointment, avoiding exposure to the aggravating factors and regular use of gloves during exposure to cold. Seventeen patients underwent classical anterior cervico-thoracic sympathectomy through supra-clavicular approach.

Table-1: Criteria to assess the severity of Raynaud's phenomena

Severe Raynaud's	Moderate Raynaud's	Mild Raynaud's
Four or more attacks in a month lasting for 15 minutes or more or onset of complications	1–3 attacks per month lasting for less than 15 minutes and vanishing without any complication	Less than one attacks per month lasting for not more than 5 minutes without any residual effect

All patients were assessed according to the criteria mentioned in Table-1 at regular intervals and data was collected on a proforma. Responders and non-responders of both groups were categorized.

Patients were followed up for 2–5 years. Chi-square test was applied to see the effectiveness of both treatment modalities. As a similar study has not been found in the literature, our study is a pilot study that does not require sample size calculation.

RESULTS

More than half of our patients were males (33), as compared to females (15). All these patients reported with typical clinical presentation of Raynaud's syndrome as a complication of frostbite after healing of wounds. Most of our patients (41) were young and below the age of 30 years.

Table -2: Age and sex distribution of 48 patients

	Total number	<20 years of age	20 to 30 years	>40 years
Male	33	7	21	5
Female	15	1	12	2

None of these patients had previous history of Raynaud's syndrome before sustaining cold injury. Over 43 (89%) patients had more than two-year history of Raynaud's syndrome which had developed after the healing of frost bite injuries. Only three patients had a shorter history of less than one year of Raynaud's syndrome.

Out of all operated patients, eleven had non-healing ulcers over the fingers. All operated patients experienced very frequent attacks of symptoms leading to prolonged hyperaemia, swelling and persistent pain in the fingers and palm of injured hand. Symptoms were triggered even by washing of hands or exposure to cold winds. Attacks lasted 6 to 12 times a week during winter, late autumn and early spring while some patients were not symptom free even during summer. Smoking was another aggravating factor.

All operated patients (94%) of group A, except one, responded well to surgical procedure in the form of remarkable reduction in the number and severity of attacks. *p*-value calculated showed significant effectiveness of cervical sympathectomy over conservatively managed patients (*p* < 0.05) (Table-3).

This improvement lasted for more than two years in these patients and they experienced very few attacks with lesser severity even during winters. Eleven (64.70%) patients were almost symptom free even after four years. Four patients (23.53%) developed Raynaud's phenomenon after two years of surgery but less in intensity and frequency in attacks compared to preoperative period, and manageable by conservative measures. Out of the operated patients, 6 were smokers. They responded very well to abstinence from smoking and experienced very few attacks after a lapse of two years of surgery and that too only during harsh winter. Out of 11 patients with non-healing ulcer on their distal fingers, 10 had

successful healing of ulcers but one patient required amputation of distal phalanx of his middle finger due to infective gangrene and required further medications and careful preventive measures to manage his severe attacks of Raynaud's phenomenon.

Only two patients (11.76%) developed complications of the surgery. One patient developed Horner's syndrome immediately after surgery, which prolonged for 4 weeks and thereafter patient recovered fully. One patient developed pneumothorax requiring intercostal intubation, and resolved without any after-effects. Rest of the patients did not develop any notable complication.

In Group-B, out of 31 conservatively managed patients, 12 patients responded well to conservative management. In these patients the duration and intensity of the Raynaud's attacks were reasonably reduced. While other 19 patient's required intensive oral and local therapy with all the preventive measure to modulate Raynaud's phenomenon especially during cold part of the year. This group also required a continuous surveillance compared to the sympathectomy group.

Table – 3: Cervical sympathectomy/conservative treatment/Favourable Response Cross-tabulation

		a Favourable Response		Total
		Yes	No	
Cervical sympathectomy	Yes	16	1	17
	No/Conservative treatment	12	19	31
Total		28	20	48

b

	Value	df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	13.868	1	0.000		

a: Computed only for a 2x2 table

b: 0 cells (0%) have expected count less than 5. Minimum expected count is 7.08.



Figure-1: Severe Raynaud's leading to ulcer of right middle finger



Figure-2: Intractable Raynaud's lead to severe ischaemia and gangrene of distal index and middle fingers.



Figure-3: Incapacitating Raynaud's healed ulcers of right fingers 2nd and 3rd of hand and contracture of 4th and 5th left fingers of after sever prolonged ischemia.

DISCUSSION

Raynaud's syndrome is characterized by periodic digital vasospasm associated with numbness, tingling and skin colour changes which vary from pallor to cyanosis to rubor. Attacks reproduced after exposure to cold and can be elicited by the ice water immersion test. The disease is more common in women than men. During the attacks pulses may or may not be present. The series of changes occur due to profound vasospasm which gradually resolves, the small amount of blood entering the capillaries rapidly desaturates and causes cyanosis. With resolution of the vasospasm arteriolar relaxation occur with resultant hyperaemia and rubor.^{2,3} The degree of severity of these tricolour phenomena varies from person to person. Hyperhidrosis may or may not be apparent and a small number of patients may have no colour changes at all. The usual episodic changes are most often precipitated by emotional stress, exposure to cold and smoking. Raynaud's usually seen in the upper limb but rarely may involve the lower limb (feet and toes).^{10,11} Maurice Raynaud in 1862 first described this condition and suggested that changes were caused by vasospasm. Allen and Brown (1932) were the first to recognize that the syndrome may occur with variety of

underlying conditions, many of which produce digital artery occlusion.¹²

Raynaud's is divided into two major groups.

a) Primary Raynaud's:

This is Raynaud's without an underlying disease or associated medical problem that could provoke vasospasm. It is the most common form of the disorder. Primary Raynaud's typically affects the digits of both hands and both feet.

b) Raynaud's disease or Secondary Raynaud's:

This is Raynaud's caused by an underlying problem. This condition occurs secondary to underlying other disease, is less common than the primary form, it's often a more complex and serious disorder.¹²⁻¹⁴

Many treatment modalities have been described for this complex problem. Most of the studies focus on conservative treatment and cervical sympathectomy has fallen out of favour due to unequivocal results in patients with primary Raynaud's syndrome. However no study evaluated the efficacy of the above mentioned treatments in patients with secondary Raynaud's disease. Our study focused on Raynaud's disease secondary to frost bite and therefore the patient group had significantly different characteristics as compared to the patients having primary Raynaud's syndrome. Our patients were predominantly male patients suffering from secondary Raynaud's phenomenon (Raynaud's syndrome) compared to most of the studies mainly designed to study female population.^{15,16}

The results of the two modalities of treatment obtained were also different as compared to the other studies.^{17,18} In our study cervical sympathectomy group had significantly better outcome as compared to the conservatively managed group. This difference may be due to etiological differences in our study and other studies.

Relapse of the disease was also not as common and not so soon as is reported in the literature.^{17,18}

CONCLUSION

Cervical Sympathectomy is an effective mean of symptomatic relief and prevention of impending complications and limb loss in Raynaud's syndrome caused by frostbite. It cures ischemic ulcers and reduces the numbers and severity of Raynaud's phenomena.

Prevention by abstinence from exposure to cold and tobacco, and management by medical means is effective in mild and some moderate cases of Raynaud's syndrome.

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