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Raynaud's Phenomenon

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Introduction

Raynaud's phenomenon (RP) is a widely prevalent clinical disorder commonly seen in outpatient settings. It is characterized by episodic vasospastic attacks of the digital arteries and arterioles that limit blood flow to the extremities, causing severe pain. Temperature changes and stress are the primary triggers that exacerbate this disease. The classic biphasic color changes of RP are pallor, cyanosis, and erythema and commonly affect the fingers and toes and more rarely, the nose, nipples, ears, lips, and penis. RP is divided into subcategories. Primary Raynaud's phenomenon (PRP) is when no underlying medical disease exists and the condition happens spontaneously. It is usually benign with mild attacks and treated with conservative measures. Secondary Raynaud's phenomenon (SRP) occurs in the context of an underlying systemic disease such as Scleroderma (Brown, 2012, p. 41). SRP patients often experience more severe attacks and require pharmacological treatments along with conservative measures. They are at greater risk of developing more severe symptoms such as digital ulcerations and gangrene (Parker, 2013, p. 23).

An increased understanding of the pathophysiology of PRP and SRP has enhanced the treatment options available for these patients. Although RP can poses problematic, debilitating symptoms for patients, healthcare professionals can help these patients learn how to self-manage their disease and live a more comfortable lifestyle. This topic was chosen to research to increase nurses' knowledge of the pathophysiology of PRP and SRP, the presentation of the disease symptoms, and the implications that the care provided can have on patient outcomes.

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Epidemiology

RP occurs more frequently in woman than in men with an onset median age of 14 and 27% of people presenting with the disease for the first time are 40 years or older (Brown, 2012, p. 42). The majority of women have PRP (85%) while 15% have SRP, whereas in men the incidence of the two types are equal (Brown, 2012, p. 42). Worldwide, the incidence of RP in adults is 4% to 20% and about 5% to 10% of all Americans (Vacca, 2014, p. 58). Non-modifiable risk factors associated with RP include being a female gender and having a family history of the disease. Modifiable risk factors include occupational use of vibrating tools, taking medications such as beta blockers or amphetamines, smoking, and underlying conditions such as atherosclerosis, hypothyroidism, polycythemia, and other hyperviscosity states.

Signs and Symptoms

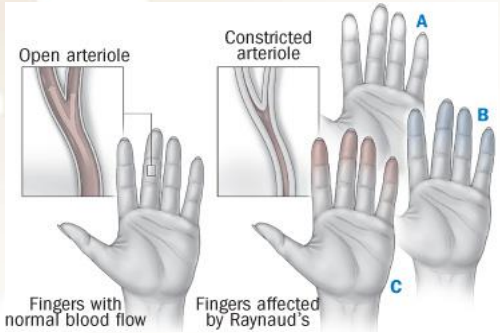
The signs and symptoms of RP depend on the severity and frequency of the vasospastic attacks. Symptoms associated with this disease include the following:

- Numbness
- Pain
- Paraesthesia

Cold temperatures or emotional stressors trigger an attack and the blood supply to the body's extremities is temporarily ceased due to vasoconstriction, causing pain and numbness (Mawdsley, 2011, p. 130). Paraesthesia occurs when the blood flow is restored to these extremities. The following signs are associated with this disease are known as the classic biphasic color changes.

- Pallor (white)
- Cyanosis (blue)
- Erythema (red)

First, a whitening (pallor) of the extremities affected (usually the fingers or toes) occurs due to ischemia from vasoconstriction, followed by a blue (cyanosis) color due to the lack of oxygen, and resolving with a red (erythema) flushed color representing reperfusion to the digits affected. Signs and symptoms usually resolve within one hour if appropriate interventions are utilized. Other symptoms associated more commonly with SRP are digital ulcerations, gangrene, skin thinning, tightening, sclerodactyly, and telangiectasias (Valdovinos & Landry, 2014, p.242). The figure below shows the classic biphasic color changes associated with RP ("Cold fingers," 2009, p. 4).



Underlying Pathophysiology

Although the pathophysiology of RP is still being explored, much has been uncovered from extensive research. The main concept is impaired perfusion to the peripheral parts of extremities triggered by cold temperatures and emotional stressors which in turn causes reversible, vasospastic attacks. In both forms of RP, the triggers lead to vasoconstriction and vasospasms. Clinically, first the affected digits become white (pallor) due to the decrease in blood flow and then progress to a blue (cyanotic) color due to deoxygenation and finally turn a flushed red (erythema) color to represent reperfusion to the affected digit after rewarming interventions have been successful (Polidoro et al., 2012, p. 532). PRP manifests with symmetric symptoms, usually a milder form in which the thumb is not affected whereas SRP may have unilateral symptoms with more severe, debilitating complications such as digital ulcerations. When the thumb is affected, this indicates SRP and that an underlying connective tissue disease is probable. The following table displays the important distinctions between PRP and SRP (Devulder et al., 2011, p. 485).

Table 2. Differences between Primary and Secondary Raynaud's Phenomenon

	Primary	Secondary
Incidence	3% to 5%	0.2%
In combination with other diseases	No	Yes
Associated with antibodies	No	Often
Dilated capillaries in nail bed	No	Often
Familial predisposition	Yes	Yes
Connective tissue disorders in family	Yes	Yes
Medicinal treatment necessary	Rarely	Often
Complications	No, rarely	Yes
Improves after some time	Yes, often	Sometimes

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During an episode there is sustained vasoconstriction with impaired vasodilation, endothelial dysfunction, and platelet activation and aggregation (Vacca, 2014, p. 58). Vasoconstriction occurs as a response to triggers in the small muscular arteries and arterioles in the digits affected. PRP is thought to be related to abnormal vasoconstriction of digital arteries and cutaneous arterioles due to a local functional defects in normal vascular responses. Evidence suggest that the defect is an increase in alpha-2 adrenergic responses in the digital and cutaneous vessels that causes arterial vasoconstriction which leads to vasospasms (Vacca, 2014, p. 58). Capillary damage prevents the endothelium from releasing vasodilating substances such as nitric oxide, prostaglandins, and neuropeptides, which further sustains vasoconstriction (Vacca, 2014, p. 58). Nitric Oxide inhibits vascular smooth muscle contraction, platelet aggregation and adhesions of platelets. With the impaired release of nitric oxide and other vasodilating substance, vasoconstriction and vasospasm persist. Furthermore, platelet activation and aggregation causes endothelial cell dysfunction and activates the release of endogenous vasoconstrictors, thus continuing to lead to sustained vasoconstriction and vasospasms (Polidoro et al., 2012, p. 532).

The underlying disease associated with SRP is thought to disrupt the normal mechanisms responsible for the control of vessel reactivity. Structural vascular changes occur with SRP. The arterial lumen narrows due to adventitial fibrosis, intimal proliferation and fibrosis, causing more severe symptoms which can lead to tissue ischemia (Sinnathurai & Schrieber, 2013, p. 478). Recurrent episodes of ischemia and reperfusion from SRP can lead to significant complications and morbidity.

Significance of Pathophysiology

Those patients suffering from RP may have a diminished quality of life due to their signs and symptoms from the disease. They can also be at high risk for developing life-altering complications, especially those with SRP. The primary goals of therapy are to improve the quality of life of patients, reduce the frequency and intensity of vasospastic attacks, and to prevent ischemic tissue injury. Understanding the mechanisms involved in the pathophysiology of this disease process is the key to providing goal directed therapy and will allow healthcare professionals to better educate their patients while providing them with a better quality of life.

Implications for Nursing Care

Advanced practice nurses (APN) must complete a full thorough history and physical exam to diagnose RP and must be able to differentiate between PRP and SRP. A careful history should include the age of onset, affected digits, and degree of severity, frequency, and symmetry of attacks. The following three screening questions should be asked to all patients with suspected RP (Wigley, 2015, p.12):

- Are your fingers usually sensitive to cold?
- Do your fingers change color when they are exposed to cold temperatures?
- Do your fingers turn white, blue, or both?

If the patient has positive answers to all three questions then a diagnosis of RP is usually made but then the APN has to inspect deeper to figure out the cause. The APN should ask the patient about potential exposures or aggravating factors such as occupational risks (frostbite, hand-arm vibration), medications (nicotine, non-selective beta-blockers), and neurological conditions (carpal tunnel syndrome). A complete physical assessment should be conducted to evaluate for other symptoms of a suggestive autoimmune disease such as fever, skin rashes, and cardiopulmonary and musculoskeletal abnormalities. The fingers and toes should especially be inspected for signs of tissue ischemia, as these are the most common digits affected. Lastly, nailfold capillaroscopy is a preferred method to distinguish between PRP and SRP. Patients with normal capillary loops around the nail bed have PRP whereas the capillary loops around the nails bed in SRP can be enlarged, disorganized, and may even hemorrhage (Brown, 2012, p. 42).

Once the diagnosis of RP has been made, either PRP or SRP, the APN must determine the severity of the disease process. It is vital to have a definitive diagnosis so that proper treatment measures can be effective. The APN must direct nursing care at preventing complications and improving the quality of life for their patients. The following are common conservative measures to prevent attacks:

- Avoid cold exposure
- Avoid rapidly changing temperatures
- Use strategies to keep the entire body warm
- Avoid cigarette smoke
- Exercise
- Manage stress with relaxation techniques and meditation
- Have knowledge on methods to help terminate an attack

Encouraging patients to develop self-management strategies helps patients establish a sense of empowerment. An important aspect of the management for RP is recognizing triggers and knowing how to prevent exacerbations. APNs need to provide support for their patients as they will be making lifestyle changes. Having a compassionate and caring healthcare team influences the patient to make healthy lifestyle changes and promotes a better well-being while decreasing stress.

Conclusion

RP is an exaggerated vascular response to cold temperatures or to emotional stress, which is characterized by the classic biphasic color changes, pallor, cyanosis, and erythema, in affected digits. In PRP, symptoms occur without an underlying disease and treatment is focused more on conservative measures due to milder attacks. SRP occurs in association with another disease such as an autoimmune disease and symptoms are more severe usually requiring conservative and pharmacological treatments. Understanding the pathophysiology of this disease process will help APNs diagnosis this phenomenon more definitively and better direct treatment goals to improving ones quality of life.

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