DEFINITION

1. Raynaud's syndrome is a disorder characterised by intermittent short-lasting inappropriate spasm of the arterioles of the distal limbs following exposure to cold or emotional stimuli.

2. The term includes on the one hand cases in which a precise cause has not been clearly defined (primary Raynaud's syndrome, Raynaud's disease, idiopathic episodic digital vasospasm) and on the other, cases where the disorder or secondary to some systemic illness or disease process (secondary Raynaud's syndrome, Raynaud's phenomenon).

CLINICAL FEATURES

3. The classic manifestation is a triphasic colour response of abrupt onset, consisting first of blanching, or pallor, due to constriction of the small arteries (vasospasm), then cyanosis due to stasis of the blood as the vasoconstriction becomes less severe, and finally the flush of reactive hyperaemia as the blood supply is restored. The affected part therefore turns white, then blue and finally red. The attack is frequently associated with numbness of the affected areas and typically lasts from a few minutes to half an hour.

4. Episodes are usually precipitated by exposure to a cool environment, although in some patients they may be triggered by emotional stress. The application of heat shortens the attack, and recovery is often accompanied by pain and throbbing.

5. In the great majority of patients, the fingers are the initial site of involvement. At first, only the tips of one or two fingers are involved in the attacks but later, colour changes may develop in additional fingers. In some 40 per cent of patients the toes are also affected and rarely, the tip of the nose or lobes of the ears.

6. In **Raynaud's disease** physical examination is normal between attacks, although minor skin changes may be noted. Thickening and tightening of the digital subcutaneous tissue (sclerodactyly) develops in 10 per cent of patients.

7. The criteria for diagnosis of primary Raynaud's syndrome include:

    7.1. Intermittent attacks of ischaemic discoloration of the extremities

    7.2. Absence of organic arterial occlusion

    7.3. Bilateral distribution

    7.4. Skin changes which are mild and localised

    7.5. Absence of signs of any signs or symptoms of systemic disease which might account for the occurrence of Raynaud's phenomenon.

    7.6. Duration of the symptoms for 2 years or longer.

8. In **secondary Raynaud's syndrome**, physical findings may reflect the underlying systemic disorder. The severity of skin changes varies considerably, and in some cases ischaemic ulceration of the tips of the fingers may occur and progress to gangrene and amputation.

AETIOLOGY

9. The patency of the digital artery depends on a favourable balance between the pressure within its lumen and the contractile forces of its muscular wall. The first is influenced by such factors as low blood pressure, arterial occlusive disorders and increased blood viscosity, the second by a complex array of factors such as increased sympathetic nervous system activity, local vascular hyper-reactivity, impaired vasodilatation mechanisms, and imbalance of circulating vasoactive hormones.

10. Although exposure to cold may precipitate an attack in vulnerable subjects, the condition is not caused by low temperatures.

11. **Raynaud's disease** Primary Raynaud's syndrome remains at present a condition of unknown aetiology, although recent research increasingly supports the view that the condition is endogenously determined. Women are affected three to five times more frequently than men and the strong familial relationship suggests a genetic link although as yet unspecified. There is no evidence that it is related to external factors.

12. Several studies have correlated Raynaud's disease with migraine headaches and variant angina, suggesting a common mechanism for the vasospasm.

13. **Secondary Raynaud's syndrome** The causes of secondary Raynaud's syndrome are numerous and varied, and include:

    13.1. **Connective tissue diseases** such as scleroderma, systemic lupus erythematosus, rheumatoid arthritis, and polymyositis.

    13.2. **Arterial occlusive diseases**; for example thromboangiitis obliterans, an inflammatory occlusive vascular disease occurring usually in men aged under 40 years. There is a clear link with smoking.

    13.3. Thoracic outlet syndrome may be included in this group. In this developmental disorder, compression of nerves and blood vessels as they pass through the neck and shoulder produce a symptom complex which includes Raynaud's phenomenon.

    13.4. Narrowing of the arteries of the upper limb due to arteriosclerosis is infrequent as an isolated occurrence, although it does occur. It is most frequent in men of over 50 years of age and women of over 60, and tends to be unilateral. It is usually part of a generalised process and coexistent arteriosclerosis of the coronary and cerebral vessels is likely.

    13.5. **Drugs and toxins** such as beta-adrenergic blocking agents eg. propranolol, and some anti-tumour antibiotics like bleomycin.

13.6. **Trauma** such as electric shock injury, and exposure to vibratory tools, grinders and sanders, where the condition forms one of the components of hand-arm vibration syndrome (HAVS). This condition is discussed in more detail in the Medical Appendix; Hand-arm Vibration Syndrome.

13.7. An episode of severe exposure to cold may induce secondary Raynaud's phenomenon; eg. frostbite may lead to vasomotor instability that can last for many years after the freezing episode, and trench foot and immersion foot may have a similar effect.

13.8. Mechanical percussive injury may cause the condition, eg. in piano playing and typing.

13.9. In the hypothenar hammer syndrome, patients develop Raynaud's phenomenon, which may be unilateral. The condition is due to hammering with the palms of the hands or such activities as karate, causing thrombosis of the ulnar artery.

13.10. **Blood disorders** Blood dyscrasias in which the normal fluid mobility of the blood is reduced, such as cold agglutinin disease and cryoglobulinaemia may also be associated with Raynaud's phenomenon, as well as the hyperviscosity syndrome that accompanies myeloproliferative diseases.

13.11. **Neurological diseases** A variety of neurological diseases, especially those involving disuse of the limb, may be associated with Raynaud's phenomenon. These include stroke, syringomyelia, intervertebral disc disease, reflex sympathetic dystrophy and poliomyelitis.

13.12. Raynaud's phenomenon occurs in 10 per cent of patients with carpal tunnel syndrome, an entrapment neuropathy caused by compression of the medial nerve as it passes through the carpal tunnel.

13.13. **Miscellaneous causes** include hypothyroidism, neoplasm and renal failure.

CONCLUSION

14. Raynaud's syndrome is characterised by episodic blanching of the extremities – most commonly the fingers. The attacks are short-lived and often triphasic in nature.

15. Raynaud's disease remains a condition of unknown aetiology. Recent evidence suggests endogenous origin, and it is likely that there is a genetic link. There is no evidence that it is linked causally to external factors.

16. Secondary Raynaud's phenomenon is attributable to a variety of causes, including toxic agents and traumatic effects, disorders of the blood and blood vessels, connective tissue diseases and diseases of the nervous system.

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