Raynaud phenomenon in children

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Abstract

Question I have several patients, mostly girls, who are living with Raynaud phenomenon. Does this condition appear in children, and what should be the course of action?

Answer Raynaud phenomenon, described in the 1860s, can present in children and even in the first decade of life. While most children will have primary Raynaud phenomenon, with no serious adverse consequences, in others it might be a sign of a pending systemic disease. Those children with a positive reaction to antinuclear antibody, specific autoantibodies associated with connective tissue disease, or nail fold capillary changes require referral to a pediatric rheumatologist and close follow-up.

Raynaud in children

It is difficult to estimate the prevalence of RP in children, as many families might perceive the colour changes as a normal response to cold exposure. One pediatric study from the United Kingdom used survey methodology with pictures, and among 720 schoolchildren, 18% of girls and 12% of boys reported a change of colour in their fingers in cold climates at least once a month, or a “numb or tingly” sensation in the fingers with cold exposure. Prevalence increased with age, especially among the girls. Another multicentre report found RP in 2.2% of children aged 0 to 10 and in 20% of those aged 10 to 20; however, there was a very wide range in the documented rate of onset among centres and between boys and girls.

Most children (about 70%) present with primary RP, and secondary RP is associated with juvenile systemic lupus erythematosus, juvenile systemic sclerosis, mixed connective tissue disease, and rarely systemic sclerosis and Sjögren syndrome. These need to be ruled out in every child with RP.

In a prospective follow-up study of 250 children and young adults with RP aged 10 to 20 (44% aged 10 to 16), nail fold capillaroscopy examination was performed, and 1 to 6 years of follow-up was available. At the end of the follow-up period, 191 (76.4%) subjects had primary RP, 27 (10.8%) had undifferentiated connective tissue disease,
and 32 (12.8%) had a specific connective tissue disease. Mean time to a form of disease was 2 years. Nonspecific capillary changes occurred in 3 out of 10 (30.0%) patients with rheumatoid arthritis, 2 out of 9 (22.2%) with systemic lupus erythematosus, 4 out of 27 (14.8%) with undifferentiated connective tissue disease, and 18 out of 191 (9.4%) with primary RP. In a pediatric series, Nigrovic et al reported retrospective chart review findings from 123 cases from Children’s Hospital of Boston; 80% were girls and 70% did not have a recognized underlying connective tissue disease. Predictive factors for an underlying condition were the presence of antinuclear antibodies and abnormal nail fold capillaries. Antiphospholipid antibodies were common but not helpful in differentiating between primary and secondary RP. Similarly, a 1989 study by Duffy et al from Toronto, Ont, reported that among 27 patients with RP (mean age at onset was 11.7 years), 33% had primary RP, 52% had a connective tissue disease, and 15% had a probable connective tissue disease. Positive reaction to antinuclear antibody and higher nail fold capillary microscopy scores were much more common in the secondary RP group.10

**Evaluation of children with RP**

Based on limited evidence in pediatrics, a European expert panel recommended testing antinuclear antibodies, more specific antibodies associated with connective tissue disease, and nail fold capillaroscopy in all children presenting with RP.11 The frequency of follow-up depends on the presence of these risk factors, with the aim of detecting evolving connective tissue disease early in high-risk individuals. Those children with a positive reaction to antinuclear antibody, specific autoantibodies, or nail fold capillary changes need a pediatric referral and close follow-up.

**Competing interests**

None declared

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**References**


**Child Health Update** is produced by the Pediatric Research in Emergency Therapeutics (PRETx) program (www.pretx.org) at the BC Children’s Hospital in Vancouver, BC. Dr Goldman is Director of the PRETx program. The mission of the PRETx program is to promote child health through evidence-based research in therapeutics in pediatric emergency medicine. Do you have questions about the effects of drugs, chemicals, radiation, or infections in children? We invite you to submit them to the PRETx program by fax at 604 875-2414; they will be addressed in future Child Health Updates. Published Child Health Updates are available on the Canadian Family Physician website (www.cfp.ca).

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**Figure 1. Pallor phase of the triphasic colour changes associated with Raynaud phenomenon**