

The St George's Classification Algorithm of Primary Lymphoedema

Dr Kristiana Gordon

Primary lymphoedema occurs as a result of a genetic predisposition causing the lymphatic system to fail to develop and/or function normally. A patient with primary lymphoedema may only have problems with swelling, but some forms of primary lymphoedema occur in association with other health problems e.g. congenital heart disease, systemic/internal lymphatic abnormalities.

Primary lymphoedema is not one disease, but the presenting feature of several distinct clinical entities. Our experience and research has led us to realise that primary lymphoedema can be divided into five different categories. Causal gene mistakes have been identified for a number of disease subtypes within the five categories. We have developed a colour-coded diagnostic pathway that describes specific primary lymphoedema phenotypes and guides the clinician on gene tests that may be available for their patient, and when to screen for associated health problems.