



Project Summary

Characterising autoantibodies in patients with Juvenile-onset scleroderma

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Scleroderma is a rare autoimmune disease that causes skin hardening called fibrosis. There are two types; localised (mainly affecting skin) and systemic (affecting skin and major organs). Up to 25% of children with localised disease have other features such as arthritis. Those with systemic disease can develop life-threatening organ involvement, including heart and lung damage.

Scleroderma affects children in different ways so it is difficult to predict what will happen as they grow. Scleroderma autoantibodies are markers detectable in the blood of patients. In adults different scleroderma autoantibodies are associated with different complications giving doctors useful information on prognosis. >95% of adult patients have a detectable autoantibody but only half of affected children. We think this is because those autoantibodies common in affected children are different from adults. We have shown this to be the case for a related condition called myositis. We know that most children with scleroderma have an autoantibody because a screening test called ANA is often positive.

Identifying key autoantibodies in juvenile scleroderma patients will provide vital prognostic information and will allow more personalised treatment and monitoring based on likely complications. A better understanding of prognosis has been identified as a key issue for affected families.