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SCIDA

Geographical and ethnic variations in disease have an intrinsic fascination. Severe combined immune deficiency (SCID) with T and B cell lymphopenia is fairly common in groups of American Indians whose main common feature is a single language, Athabascan. They are, in the main, members of three tribes, Navajo and Apache in southwest USA and the Dogrib in northwest Canada. Their ancestors were hunter gatherers in the northwest some of whom began to migrate south in about the eighth century AD, the Navajo and Apache tribes forming in the 17th century. In these people the incidence of SCID is about 1 in 2000 births whereas in the US population as a whole it is around 1–2 per million live births. The SCID in Athabascan speakers (SCIDA) is autosomal recessive, the gene being on chromosome 10p. A peculiarity of this form seems to be presentation with oral and genital ulcers.

Over a period of 8½ years 12 children with SCIDA presented to a bone marrow transplant unit in San Francisco (Pearl C Kwong and colleagues. *Archives of Dermatology* 1999;**135**:927–31). Ten developed oral or genital ulcers or both before the age of 3 months. All had severe T and B cell lymphopenia. None of 21 children from non-Athabascan speaking families who presented with SCID during the same period had orogenital ulcers. The ulcers healed after successful marrow transplantation, T cell function apparently being more important than B cell.

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