Prkdc<sup>scid</sup>
Severe combined immuno deficiency

**Origin:**
In 1980 at Fox Chase Cancer Centre, this mutation was discovered by Bosma (Bosma et al., 1983). This mutation occurred in the C.B-17/Icr strain, an Igh congenic partner of BALB/cAnIcr differing from it only by a portion of chromosome 12 that was derived from the C57BL/Ka strain.

- **BALB/cJHan<sup>TM</sup>Hsd-Prkdc<sup>scid</sup>**
  In 1989, to Central Institute for Laboratory Animal Breeding, Hannover, Germany. In 1994, to Harlan UK through acquisition.

- **C3H.C-Prkdc<sup>scid</sup>/IcrSmnHsd**
  In the C3H.C-Prkdc<sup>scid</sup> is the donor strain C.BKa-Igh<sup>b</sup>/IcrSmn (C.B-17) and the background strain C3H/HeSnSmn. To Harlan Sprague Dawley, Inc.

- **C.B-17/IcrHan<sup>TM</sup>Hsd- Prkdc<sup>scid</sup>**
  In 1989, to Central Institute for Laboratory Animal Breeding, Hannover, Germany. In 1994, to Harlan UK through acquisition.

- **HsdIcr:Ha(ICR)- Prkdc<sup>scid</sup>**
  At the Institute for Cancer Research, Philadelphia, the scid gene was transferred to the ‘Swiss-Webster’ stock IcrHa:CRI. To Harlan Sprague Dawley, Inc.

**Research Applications**
Immunology, xenografts, *Pneumocystis carinii* infection.

**Characteristics:**

- **Anatomy**
  Scid mice have no abnormal external characteristics. The lymph nodes and thymus are abnormally small, as is the spleen of most animals. The thymus consists of a rudimentary medulla with a cortex. Spleen and lymph node follicles are virtually devoid of lymphocytes. All of these lymphoid organs consist primarily of vascularized supportive tissue with variable numbers of fibroblasts, histiocytes, and macrophages. Bone marrow, although lacking lymphocytes and plasma cells, appears otherwise to be morphologically normal (Bosma et al., 1983; Custer et al., 1985; Dagnæs-Hansen et al., 1991).

- **Husbandry**
  Homozygous scid mice readily succumb to microbial infections because of their lack of an immune system and must be maintained in isolators, micro-isolators, laminar flow cabinets or pathogen free environment. Under these conditions their lifespan can be 9-12 months of age (ILAR, 1989).

- **Genetics**
  Scid is an autosomal recessive mutation that occurred spontaneously in the CB-17 congenic strain. In this congenic strain the inbred BALB/c strain carries the
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immunoglobulin heavy-chain allele *Igh-1<sup>b</sup>* of the C57BL/Ka strain. The *scid* locus has been mapped to the centromeric end of chromosome 16 (Bosma *et al*, 1989).

Mutant stock gene - *scid*. See parental strains for additional genes.

- **Immunology**
  
  T-cell development in *scid* mice equals the functional stage of T-cell in 14 day foetuses. Most thymocytes are negative for specific T-cell antigens (CD3, CD4<sup>-</sup>, CD8<sup>-</sup>), but express low levels of CD2 (Habu *et al*, 1987; Hardy *et al*, 1989). The CD3 has an obligate co-expression with the T-cell antigen receptor and is found in the mature T-cell lineage. A large fraction of the cells in the *scid* thymus expresses the smaller chain of the IL-2 receptor, which also is a characteristic of immature CD4<sup>-</sup>, CD8<sup>-</sup> thymocytes in normal mice. Mice with the *scid* mutation lack dendritic Thy-1<sup>+</sup> epidermal cells (Nixon-Fulton *et al*, 1987).

  *Scid* mice cannot reject allogeneic grafts or produce antibodies to common laboratory antigens, and their spleen cells do not proliferate in response to T- or B-cell-specific mitogens. Fluorochrome-conjugated antibody reagents that specifically stain B and pre-B cells fail to detect such cells in the spleen and bone marrow (Bosma *et al*, 1983; Dorshkind *et al*, 1984).

  *Scid* mice have the recessive mutation 'severe combined immunodeficiency', causing greatly reduced numbers of T cells and B cells. Homozygotes are deficient in both B and T cell function. Consequently, these mice are deficient in both humoral and cell-mediated immune function and lack detecting levels of circulating immunoglobulins. Most homozygotes have no detectable IgM, IgG1, IgG2a, IgG2b, IgG3, or IgA, but a few have low levels of one to three of these immunoglobulin isotypes. *Scid* mice with serum Ig levels greater than 1 µg/ml are considered 'leaky'. Low level of "leakiness" when the *scid* mutation is maintained on the C3H background (contrast C.B-17) (Nonoyama *et al*, 1993).

- **Infection**

  The *scid* mouse is a model for the analysis of Lyme arthritis and carditis (Schaible *et al*, 1989).

- **Life-span and Spontaneous Disease**

  Under SPF conditions, homozygous *scid* mice may survive a year or more. About 15% of all *scid* mice spontaneously develop thymic lymphomas (Custer *et al*, 1985) that contains rearranged T-cell receptor gamma and T-cell receptor beta alleles and express T-cell specific cell surface antigens.

- **Miscellaneous**

  The *scid* mice have clearly demonstrated to be a valuable model for the study of the immune system, especially the development of the gene rearrangement for the development of functionally immune globulins and T-cell receptors, but also for the study of reconstitution, lymphocyte trafficking and acquired immune function resulting from transfer of specific lymphoid populations (Bosma *et al*, 1991; Hilbert *et al*, 1991). Characteristics of *scid* mice have been described by ILAR (1989) and Lyon *et al* (1996).

- **Reproduction**

  Homozygous *scid* mice are fertile and can be bred without difficulty, although the average litter size (4-6) is smaller than that of the congenic C.B-17 strain (6-9).
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- **Xenografts**
The *scid* mouse has been used as a recipient for transplanted human tumours (Reddy *et al.*, 1987), human foetal lymphoid tissues (McCune *et al.*, 1988), and peripheral blood lymphocytes (Mosier *et al.*, 1988). Both latter xenotransplanted *scid* mice (scid-hu) have been recognized as an animal modal for HIV infection and antiviral testing (Bonyhadi *et al.*, 1991; Mosier 1991; Hesselton *et al.*, 1991).

**References:**
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