

## Rebecca H. Buckley, MD

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### BIOGRAPHICAL SKETCH

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NAME Buckley, Rebecca H.		POSITION TITLE J. Buren Sidbury Professor of Pediatrics Professor of Immunology	
EDUCATION/TRAINING <i>(Begin with baccalaureate or other initial professional education, such as nursing, and include postdoctoral training.)</i>			
INSTITUTION AND LOCATION	DEGREE <i>(if applicable)</i>	YEAR(s)	FIELD OF STUDY
UNC School of Medicine, Chapel Hill, NC	A.B.	1954	Pre-Medicine
Duke University Medical Center	M.D.	1958	Medicine
Duke University Medical Center	-	1958-1961	Pediatric Residency
Duke University Medical Center	-	1961-1963	Allergy Fellowship
Duke University Medical Center	-	1963-1965	Immunology Fellowship

#### A. Positions and Honors.

Research Associate, Immunology	Duke Medical Center	7/01/65-10/31/68
Asst. Professor of Pediatrics	Duke Medical Center	11/01/68-06/30/72
Asst. Professor of Immunology	Duke Medical Center	7/01/69-06/30/72
Assoc. Professor of Pediatrics	Duke Medical Center	7/01/72-04/30/76
Assoc. Professor of Immunology	Duke Medical Center	7/01/72-06/30/79
Professor of Pediatrics	Duke Medical Center	5/01/76-06/30/79
Professor of Immunology	Duke Medical Center	7/01/79-present
J.B. Sidbury Professor, Pediatrics	Duke Medical Center	7/01/79-present
Chief, Pediatric Allergy/Immunology	Duke Medical Center	7/01/74-06/30/03

#### Apha Omega Alpha

Scholar/Teacher of the Year Award, Duke University 1984

Allergic Diseases Academic Award, NIAID, 1974-1979

National Institutes of Health Merit Research Award, 1987-1997

2nd Annual Award for Excellence in Clinical Research, GCRCP of NCRR, 5/7/90

Medical College of Pennsylvania 1991 National Board Award, April 1991

Distinguished Teacher and Faculty Awards, Duke Medical Alumni Association, November 1993 and November 1998

Outstanding Achievement Award, Immune Deficiency Foundation, November 1994

American Acad. of Allergy & Immunology, Fellow (Exec. Com. 1975-82; President 1979-80, Honorary Fellow, 1999)

Allergy and Immunology Research Committee, NIAID, NIH 1972-75

Immunological Sciences Study Section, NIH 1976-80; Chairman 1979-80

National Advisory Council, NIAID, 1985-86

Board of Scientific Counselors, NIAID, 1991-1996  
Director, American Board of Allergy and Immunology 1971-73, 1982-87, President 1983-84  
American Pediatric Society, President 1999-2000  
Chairman, Medical Advisory Committee, Immune Deficiency Foundation, 2003  
Elected to the Institute of Medicine, National Academy of Sciences, 2003

**Selected peer-reviewed publications (in chronological order).** From 275 manuscripts:

1. **Buckley RH**, Schiff SE, Sampson HA, Schiff RI, Markert ML, Knutsen AP, Hershfield MS, Huang AT, Mickey GH, Ward FE: Development of immunity in human severe primary T cell deficiency following haploidentical bone marrow stem cell transplantation. *J Immunol* 136:2398-2407, 1986.
2. Schiff SE, **Buckley RH**: Modified responses to recipient and donor B cells by genetically donor T cells from human haploidentical chimeras. *J Immunol* 138:2088-2094, 1987.
3. Roberts JL, Volkman DJ, **Buckley RH**: Modified major histocompatibility complex (MHC) restriction of donor-origin T cells in humans with severe combined immunodeficiency (SCID) transplanted with haploidentical bone marrow stem cells. *J Immunol* 143:1575-1579, 1989.
4. Gaines AD, Schiff SE, **Buckley RH**: Donor type natural killer cells after haploidentical T cell depleted bone marrow stem cell transplantation in a patient with adenosine deaminase deficient severe combined immunodeficiency. *Clin Immunol Immunopathol* 60:299-304, 1991.6.
5. Russell SM, Tayebi N, Nakajima H, Riedy MC, Roberts JL, Aman MJ, Migone T-S, Noguchi M, Markert ML, **Buckley RH**, O'Shea JJ, Leonard WJ: Mutation of Jak3 in a patient with SCID: Essential role of Jak3 in lymphoid development. *Science* 270:797-799, 1995.
6. **Buckley RH**, Schiff RI, Schiff SE, Markert ML, Williams LW, Roberts JL, Puck JM: Human severe combined immunodeficiency (SCID): genetic, phenotypic and functional diversity in 108 infants. *J Pediatrics* 129:378-387, 1997.
7. Puck JM, Pepper AE, Henthorn PS, Candotti F, Isakov J, Whitwam T, Conley ME, Fischer RE, Rosenblatt HM, Small TN, **Buckley RH**: Mutation analysis of IL2RG in human X-linked severe combined immunodeficiency. *Blood* 89:1968-1977, 1997.
8. Jabara HH, **Buckley RH**, Roberts JL, Lefranc G, Loiselet J, Khalil G, Geha RS: Role of Jak3 in CD40-mediated signaling. *Blood* 92: 2435-40, 1998.
9. Puel A, Ziegler SF, **Buckley RH**, Leonard WJ: Defective IL-7R expression in T(-)B(+)NK(+) severe combined immunodeficiency. *Nature Genetics*, 20:394-397, 1998.
10. Grimbacher B, Schaffer AA, Holland SM, Davis J, Gallin JI, Malech HL, Atkinson TP, Belohradsky BH, **Buckley RH**, Cossu F, Espanol T, Garty B, Matamoros N, Myers LA, Nelson RP, Ochs HD, Renner ED, Wellinghausen N, Puck JM: Genetic linkage of Hyper IgE syndrome to chromosome 4. *Am J Hum Genet* 65:735-744, 1999.
11. **Buckley RH**, Schiff SE, Schiff RI, Markert ML, Williams LW, Roberts JL, Myers LA, Ward FE: Hematopoietic stem-cell transplantation for the treatment of severe combined immunodeficiency. *New England Journal of Medicine* 340: 508-516, 1999.
12. Patel DD, Gooding ME, Parrott RE, Curtis KM, Haynes BF, **Buckley RH**: Thymic function after hematopoietic stem-cell transplantation for the treatment of severe combined immunodeficiency. *New England Journal of Medicine* 342:1325-1332, 2000.
13. **Buckley RH**: Primary immunodeficiency diseases affecting lymphocytes. *New England Journal of Medicine*, 343: 1313-1324, 2000.
14. Minegishi Y, Lavoie A, Cunningham-Rundles C, Bedard P-M, Hebert J, Cote L, Dan K, Sedlak D, **Buckley RH**, Fischer A, Durandy A and Conley ME: Mutations in activation-induced cytidine deaminase in patients with Hyper IgM syndrome. *Clin Immunol* 3: 203-210, 2000.
15. Morra M, Silander O, Calpe-Flores S, Choi M, Oettgen H, Myers L, Etzioni A, **Buckley RH**, and Terhorst C: Alterations of the X-linked lymphoproliferative disease gene SH2D1A in the Common

Variable Immunodeficiency syndrome. *Blood* 98: 1321-1325, 2001.

16. Myers LA, Patel DD, Puck JM, **Buckley RH**: Hematopoietic stem cell transplantation for severe combined immunodeficiency (SCID) in the neonatal period leads to superior thymic output and improved survival. *Blood* 99: 872-878, 2002.

17. Sarzotti M, Patel DD, Li X, Ozaki DA, Cao S, Parrott RE, Coyne K, **Buckley RH**: T cell repertoire development in humans with severe combined immunodeficiency (SCID) after non-ablative allogeneic marrow transplantation. *J Immunol*, 170: 2711-2718, 2003.

18. Roberts JL, Lengi A, Brown SM, Chen M, Zhou Y-J, O'Shea JJ, **Buckley RH**: Janus Kinase 3 (JAK3) Deficiency: Clinical, Immunologic and Molecular Analyses of 10 Patients and Outcomes of Stem Cell Transplantation. *Blood* 103: March 15, 2004.

19. Hale LP, **Buckley RH**, Puck JM, Patel DD: Abnormal development of thymic dendritic and epithelial cells in X-linked severe combined immunodeficiency. *Clinical Immunology* 110: 63-70, 2004.

20. Kalman L, Lindegren ML, Kobrynski L, Vogt R, Hannon H, Howard JT, **Buckley RH**: Mutations in selected genes required for T cell development: *IL7R*, *CD45*, *IL2R* gamma chain, *JAK3*, *RAG1*, *RAG2*, *ARTEMIS* and *ADA* and Severe Combined Immunodeficiency. *Genetics in Medicine* 6: 16-26, 2004.

21. **Buckley RH**: Molecular defects in human severe combined immunodeficiency and approaches to immune reconstitution. *Annual Review of Immunology* 55: 20.1-20.31, 2004.

**C. Research Support.** List selected ongoing or completed (during the last three years) research projects (federal and non-federal support). Begin with the projects that are most relevant to the research proposed in this application. Briefly indicate the overall goals of the projects and responsibilities of principal investigator identified above.

**52 R01 AI042951-06A1** (Buckley) 05/01/04-04/30/09 30%

NIAID \$248,173

Development and Persistence of Immunity in SCID Chimeras

Major Goals: To investigate the ontogeny and persistence of T, B, and NK cell function in infants and children with well-characterized human severe combined immunodeficiency who are or who become haploidentical stem cell chimeras. Ov

Overlap: None

**1R01 AI47605-02** (Buckley) 09/01/99-08/31/04 30%

NIAID \$177,537

Mechanisms of Allogeneic Stem Cell Education in SCID

Major Goals: To investigate the mechanisms of allogeneic stem cell development into mature and functioning T and B cells in humans with severe combined immunodeficiency (SCID) who have become chimeric following receipt of T cell-depleted allogeneic bone marrow stem cells without pre-transplant cytoreduction or post-transplant GVHD prophylaxis.

Overlap: None

**5T32 AI07062-24** (Buckley) 09/1/00-6/30/04 20%

NIAID \$123,444

Research Training in Allergy and Clinical Immunology

Major Goals: The primary purpose of this program has been and will continue to be the training of young

physicians with strong academic potential in such a way that they will qualify for full time medical school faculty positions as allergists and/or clinical immunologists. The training will place strong emphasis on research training and acquisition of basic immunologic knowledge. These goals will be accomplished through a program which included: 1) formal courses in basic immunology and other related disciplines, 2) precepted research training, and 3) limited clinical exposure to a variety of allergic and immunologic problems so that experience can be gained in the evaluation of patients with these disorders.

Overlap: None