Pediat. Res. *11:* 65 (1977)

I. Introduction

RUSSELL J. BLATTNER

Department of Pediatrics, Baylor College of Medicine and Texas Children's Hospital, Houston, Texas, USA

Infants born with SCID rarely survive the first year of life because of their severely impaired ability to resist infection. The most effective method for correcting the deficiency in the human has been bone marrow transplantation. Bone marrow transplants from sibling donors matched at the HLA and MLC loci have been successful in restoring both humoral and cellular immunity in patients with SCID. Graft vs. host (GVH) disease has been a serious and frequently fatal complication (8, 55) and has necessitated refinements in this and other procedures for successful reconstitution of immune deficient patients. Yet in all cases of SCID there is a danger of death from infection before reconstitution can be achieved. Moreover, in some cases where reconstitution is successfully established without serious damage from GVH, death may occur from a pre-existing viral, protozoan, or bacterial infection. An approach to this problem was attempted in two children in Germany, nonidentical twins with a primary combined immune deficiency, lymphopenic hypo-γ-globulinemia (49). After diagnosis of their deficiency they were decontaminated and, at age 6 weeks, were placed in protective isolation to prevent reinfection. They survived in the isolators and, surprisingly, showed a slow maturation of the immune responses so that, at 30 and 32 months, respectively, they could be released into the unprotected environment.

The case to be presented here is that of a boy with SCID who has been reared from birth in strict reverse isolation to the age of 4 years. The possibility of an immune deficiency disorder was anticipated before birth in this case because of a brother who had SCID. The child was placed in the isolator at birth to prevent infection until a bone marrow transplant could be made. Unfortunately, no nonreactive donor for bone marrow transplantation has been found for him. Therefore, he has been maintained in a simplified and specially constructed plastic isolator system. This is a completely closed system which the child never leaves and which the attendants never enter but which allows the child and attendants to see each other clearly at all times.

The survival and continuing excellent health of a child with SCID for such a long time and under such unusual circumstances has provided a unique opportunity for observation and study. A number of interesting and unexpected findings have led to more intensive research into various aspects of his developmental progress as well as into the nature of his immunologic defect. The present report, therefore, includes a comprehensive survey of his care and development, along with related research studies during his life to the age of 4 years under the conditions of strict reverse isolation.

 Printed in U.S.A.

Pediat. Res. *11:* 56 (1977)

II. Family Background, Early History, and Diagnosis

JOHN R. MONTGOMERY, MARY ANN SOUTH, RAPHAEL WILSON, AND ROSALINDA SORIANO

Department of Pediatrics, Baylor College of Medicine and Texas Children's Hospital, Houston, Texas, USA

The parents are from the upper middle income bracket and are in good mental and physical health. They have had two other children, including a girl who is 3 years older than the patient and who appears to be normal in every respect, and a boy who died at 7 months with SCID. There is no evidence of immune deficiency in either parent or in the genetic background on the maternal side. Three male children of a brother of the mother's father may have had immune deficiencies. Two had been treated during childhood for γ-globulin deficiency but the treatment had been discontinued and the two grew to be teenagers with no apparent problems. Another died suddenly at 8 months, reportedly with "Asian influenza." At autopsy, however, lymph nodes in this infant appeared to be normal with abundant lymphocytes.

The deceased older brother of the patient had been under the care of one of us (Dr. J. R. Montgomery) and had been diagnosed to have SCID in infancy. This first son had received a