INTRODUCTION

Background: Children with Down syndrome (DS) are at increased risk of developing acute lymphocytic leukemia (ALL) and acute myeloid leukemia (AML). Leukemias occurring among DS children have been studied extensively, however, these studies have not adjusted for confounding variables.

OBJECTIVES: The purpose of this study was to use population-based cancer data to determine the characteristics and survival of children with acute leukemia according to the presence of DS or other birth defects.

RESULTS

- **ALL and AML cases diagnosed between 1983 and 2012 among children with a birth year of 1983 - 2012 and age of 0 - 14 years old were obtained from the New York State Congenital Malformations Registry (NYSCMR).**

- **Data on children with birth defects were obtained from the New York State Congenital Malformations Registry (NYSCMR).** Children with at least one reportable major birth defect and born during the same time period as the cancer cases were selected.

- **The birth defect status of cancer cases (DS, other birth defects, or no birth defect) was determined by linking the cancer case file with the birth defect data using CDC’s UniPro software.**

- **Associations between birth defect status and demographic characteristics, including age at cancer diagnosis, race, gender, and ethnicity, were evaluated using contingency table analysis. Chi-square statistics or Fisher’s exact test (for small case counts) was used to determine the significance of the association.**

- **10-year overall survival was calculated by birth defect status and other potential prognostic factors (age, gender, race, ethnicity, and year of diagnosis for ALL and AML, respectively, using the actuarial estimation method). The Log-rank test was used to evaluate the survival differences among comparison groups.**

- **Con proportional regression analysis was also performed to assess the effect of the birth defect status on survival adjusting for confounding variables.**

RESULTS (CONTINUED)

- **Among DS children diagnosed with ALL:**

  - 1.0% had DS, 3.9% had other birth defects, and 95.1% had no birth defect;
  - Age at ALL diagnosis was significantly associated with birth defect status, and the most notable increase in age at diagnosis occurred for younger children. 34.2% of ALL were diagnosed during infancy (≤ 1 year old) for children with other birth defects, compared to 3.3% for children without birth defects. None of the children with DS were diagnosed within one year of age (Table 1);
  - 10-year survival rates were consistently high, ranging from 85.1% for children with DS to 80.6% for children with no birth defect. There were no significant survival differences among the three groups (Fig. 1A, Table 2).

- **ALL children diagnosed during infancy had the worst survival outcome compared to other age groups.** Being black or Hispanic was also associated with poorer survival. Survival rates have been improving over time (Table 2).

ACKNOWLEDGEMENTS

This work was supported in part by the Centers for Disease Control and Prevention and the New York State Department of Health through the National Program of Cancer Registries.