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**POPULATION PID**

**FREQUENCY AND A SPECTRUM OF MALIGNANCIES IN 120 EAST SLAVIC PATIENTS WITH NIJMEGEN BREAKAGE SYNDROME**

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**Background:**

Nijmegen Breakage Syndrome (NBS) is a rare DNA repair disorder with high predisposition to lymphoid malignancies, which are the main cause of death in patients with NBS.

**Methods:**

We aimed to present frequency and a profile of malignancies in a large cohort of NBS patients of East Slavic origin.

**Results:**

Clinical data of 120 NBS patients from Belarus (n=20), Ukraine (n=50), Russia (n=45) and Latvia (n=5) (Russian origin) were collected. 78 out of 120 (65%) NBS pts (median age 11 years) remained alive at the time of analysis. Fifty-five malignancies were diagnosed in 53 (44.2%) NBS patients, among them 38% (20 out of 53) are alive. The first episode of malignancy occurred at a median age of 9 years (range, 2–21 years). Non-Hodgkin lymphoma (NHL) was diagnosed in 34(64%) cases, mostly (62.1%) of B-cell origin (diffuse large B-cell lymphoma (DLBCL)-14 pts, Burkitt lymphoma in 1, B-NHL (undefined) in 3). Lymphoblastic lymphoma was diagnosed in 9 pts T-cell in 8, undefined – 1), peripheral T-cell lymphoma, NOS - 2 pts., anaplastic large cell lymphoma-1. ALL was diagnosed in 13, AML – 1, Hodgkin lymphoma in 2, and lymphoma not-specified in 3. There were 4 cases of solid tumors: rhabdomyosarcoma in 2, hepatoblastoma in 1, CNS tumor in 1. Two pts with DLBCL developed second malignancy (NHL and medulloblastoma, 4 and 11 years after the first tumor diagnosis).

**Conclusion:**