OC52 A RETROSPECTIVE STUDY OF MYELOID LEUKAEMIA IN CHILDREN WITH DOWN SYNDROME IN IRELAND

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Background Haematological abnormalities are common in children with trisomy 21. These children have a remarkably high risk of acute leukaemia. The incidence of acute myeloid leukaemia is 150 fold greater in young children with DS compared to children of the same age without DS. Acute Megakaryoblastic Leukaemia (AMKL) is a subtype of myeloid leukaemia and is the most common leukaemia type in children with DS under 4 years of age. AMKL is often preceded by a transient neonatal pre-leukaemic syndrome, called Transient Myeloproliferative Disorder (TMD). Although TMD often spontaneously resolves, 20–30% of these patients subsequently develop AMKL within the first 4 years of life.

Aims To perform a retrospective consecutive national audit of all documented cases of childhood TMD and AMKL-DS from 1990–2018 at the National Paediatric Haematology/Oncology Centre, Our Lady’s Children’s Hospital Crumlin (OLCHC) Ireland. We also aimed to compare our demographic and outcome findings with the recent medical literature and make recommendations for future research.

Methods Charts of the patients with a diagnosis of AMKL and treated consecutively at (OLCHC) between 1990 – 2018 were sourced from medical records. Charts and the hospital database were reviewed and information including date and age of diagnosis of AML, comorbidities, previous history of TMD, treatment regime and outcome was noted. The Hospital Patient Administration System (PAS) was interrogated for additional information such as percentage blasts in the bone marrow and blood count values at time of diagnosis of AMKL. Cytogenetic information was obtained by accessing a dedicated database. Kaplan-Meier survival curves were constructed.

Results Twenty-seven patients with AMKL-DS were treated at OLCHC. A prior neonatal diagnosis of TMD was described in 10 patients (37%). Patients had a low platelet count (median 34 × 10⁹/L) at presentation. Nineteen patients (70%) are alive and well, in complete remission, at a median follow up of 11.4 years. Overall survival (OS) of this cohort has risen from 54% from those treated between the years 1990 – 2004 (n = 13) to 93% for those treated between the years 2005 – 2018 (n = 14).

Conclusion High cure rates are observed in AMKL-DS using current polychemotherapy protocols. However, in general children with DS are more prone to the cytotoxic effects of chemotherapy, which can lead to significant treatment related mortality. The finding of a low platelet count at time of diagnosis is in keeping with the knowledge that AMKL-DS is a malignancy of platelet progenitor cells.