

EORTC study suggests detecting ERG gene deletion useful for risk stratification in childhood ALL

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Results of EORTC trial 58951 suggest that detecting ERG gene deletion at diagnosis of childhood B-cell precursor (BCP) acute lymphoblastic leukemia (ALL) would be useful for risk stratification. The study, published in *Leukemia* showed that patients with the ERG gene deletion had a very good outcome with an 8-year event-free survival of 86.4% and an overall survival of 95.6%.

ALL is the most common childhood malignancy, but it is characterized by a number of recurring genetic alterations. These alterations, each with a specific gene expression profile, can influence response to treatment. For example, high hyperdiploidy and the chromosomal translocation t(12;21)/ETV6–RUNX1 are the most prevalent alterations in young children and are associated with good treatment response and outcome. On the other hand, t(9;22)/BCR–ABL1, rearrangements of the MLL gene, low hypodiploidy, intrachromosomal amplification of chromosome 21 (iAMP21) are all associated with a high risk of relapse. In addition, IKZF1 gene deletion has been recently described as a strong marker of poor outcome.

Dr. Emmanuelle Clappier of the Hematology University Institute, St-Louis and Robert Debré Hospitals in Paris and lead author of this EORTC publication says, "The genetic basis of BCP-ALL is still unknown for a significant proportion of cases, and consequently outcome is unpredictable at the time of [diagnosis](#). This is especially true

for older children and adolescents, more than half of whom display no classifying genetic alteration. There is a clear need for new biological markers to assist in making treatment decisions and improve outcome for these patients."

A genomic deletion in the ERG gene was identified by array-CGH analysis in selected patients. Then an independent non-selected cohort of 897 children aged 1-17 years and treated for BCP-ALL in the EORTC 58951 trial between December 1998 and July 2008 was screened for ERG gene deletions. ERG gene deletion was found in 3.2% of the patients (29 out of the 897 patients) and was associated with higher age (median age 7.0 years versus 4.0 years, $P=0.004$) and frequent IKZF1 $\Delta 4-7$ deletions (37.9% versus 5.3% in the remaining patients, P

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