

Uncovering the pathology of a rare pediatric leukemia

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A team of scientists has demonstrated the mechanism by which ETO2-GLIS2, a gene fusion, promotes the development of an aggressive form of pediatric leukemia. The findings, published in *Cancer Cell*, also reveal an opportunity for the development of therapeutics.

The study was co-authored by John Crispino, PhD, the Robert I. Lurie, MD, and Lora S. Lurie Professor of Medicine in the Division of Hematology and Oncology.

Acute megakaryoblastic [leukemia](#) (AMKL), a rare type of blood cancer predominantly found in children, has two major pediatric subgroups: AMKL in patients with Down syndrome and those without. While the disease in those with Down syndrome (DS) is relatively well-defined and carries a good prognosis, non-DS AMKL is much less well-understood.

Recently, scientists discovered that a gene fusion called ETO2-GLIS2—produced by an inversion on chromosome 16—is present in 20 to 30 percent of cases of non-DS AMKL, and is associated with a very poor patient prognosis. But up until now, it was unclear exactly how this [gene fusion](#) blocks normal cell differentiation, a hallmark of leukemia.

In the current study, the scientists illustrated how ETO2-GLIS2 induces an irregular transcription network that underlies AMKL. They further demonstrated that expression of a peptide that inhibits ETO2-GLIS2 oligomerization could release the differentiation block—insights which

could inform the development of novel therapeutics.

"Acute megakaryoblastic leukemia is a devastating [blood cancer](#) that requires new targeted and efficacious therapies," Crispino said. "The discovery of the mechanism by which ETO2-GLIS2 fusion promotes leukemia provides important new insights into ways to target these malignant cells."

More information: Cécile Thirant et al. ETO2-GLIS2 Hijacks Transcriptional Complexes to Drive Cellular Identity and Self-Renewal in Pediatric Acute Megakaryoblastic Leukemia, *Cancer Cell* (2017). [DOI: 10.1016/j.ccell.2017.02.006](https://doi.org/10.1016/j.ccell.2017.02.006)

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