

## PROF. DR. FELIX NIGGLI

University Children's Hospital  
Department of Pediatric Oncology  
Steinwiesstrasse 75, 8032 Zürich

[felix.niggli@kispi.uzh.ch](mailto:felix.niggli@kispi.uzh.ch)  
[www.kispi.uzh.ch](http://www.kispi.uzh.ch)



**KEYWORDS** — Sarcoma, interactome, oncogenic drivers, kinase inhibitors

## SUMMARY & MISSION STATEMENT

The main field of our research activity is the elucidation of the molecular mechanisms underlying development of paediatric sarcomas with special emphasis on the role of oncogenic fusion proteins generated by chromosomal translocations. Novel insights into these mechanisms are used in translational approaches to advance diagnosis and treatment in paediatric patients, carried out in close collaboration with clinical studies.

## OVERVIEW

We use mainly two tumour models in our laboratory, namely, the paediatric soft tissue sarcoma rhabdomyosarcoma as well as a paediatric bone tumour, Ewing's sarcoma, which are both characterized by specific chromosomal translocations. In Ewing sarcoma (ES) we address possible regulatory mechanisms to modulate the activity of the specific oncogenic event, EWS/FLI1. In the focus of our projects are tumour-specific oncogenic transcription factors which we regard as very important novel therapeutic targets. We characterize the biochemistry of the fusion protein and the post transcriptional mechanisms regulating the activity of the fusion protein. We aim to characterize early tumorigenic events by developing a novel ES tumour model based on reprogramming of engineered fibroblast.

## SELECTED CANCER RELATED PUBLICATIONS

Proteasomal Degradation of the EWS-FLI1 Fusion Protein Is Regulated by a Single Lysine Residue. Gierisch ME, Pfistner F, Lopez-Garcia LA, Harder L, Schäfer BW, Niggli FK. **J Biol Chem.** 2016 Dec 23;291(52):26922-26933

Targeting the EWS-ETS transcriptional program by BET bromodomain inhibition in Ewing sarcoma. Hensel T, Giorgi C, Schmidt O, Calzada-Wack J, Neff F, Buch T, Niggli FK, Schäfer BW, Burdach S, Richter GH. **Oncotarget.** 2016 Jan 12;7(2):1451-63

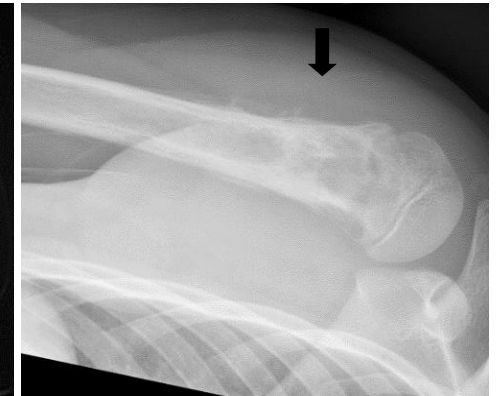
Giorgi C, Boro A, Rechfeld F, Lopez-Garcia LA, Gierisch ME, Schäfer BW, Niggli FK. PI3K/AKT signaling modulates transcriptional expression of EWS/FLI1 through specificity protein 1. **Oncotarget.** 2015 Oct 6;6(30):28895-910

Results for patients with sarcoma not otherwise specified and other diagnoses than Ewing sarcoma treated according to the Euro-EWING 99 trial. Frank JA, Ranft A, Paulussen M, Juergens H, Kruseova J, Bauer S, Niggli E, Reichardt P, Dirksen U. **Pediatr Blood Cancer.** 2017 Apr 24. doi: 10.1002/pbc.26524

FGFR4 signaling couples to Bim and not Bmf to discriminate subsets of alveolar rhabdomyosarcoma cells. Wachtel M, Rakic J, Okoniewski M, Bode P, Niggli E, Schäfer BW. **Int J Cancer.** 2014 Oct 1;135(7):1543-52.



Rhabdomyosarcoma of the bladder



Ewing sarcoma of the humerus bone