

# ZytoLight® SPEC BCOR Dual Color Break Apart Probe



## Background

The ZytoLight® SPEC BCOR Dual Color Break Apart Probe is designed to detect rearrangements involving the chromosomal region Xp11.4 harboring the BCOR (BCL6 corepressor, a.k.a. KIAA1575) gene.

In the 2020 WHO classification of soft tissue and bone tumors, BCOR-rearranged sarcoma is recognized as a distinct entity due to particular morphological, immunohistochemical, and molecular features and differing clinical outcomes compared to other undifferentiated sarcomas.

A fusion between BCOR and CCNB3 can be found in about 60% of all BCOR-rearranged sarcomas. The BCOR-CCNB3 fusion results from an X-chromosomal paracentric inversion. In vitro studies suggest that the BCOR-CCNB3 fusion protein is oncogenic and drives proliferation in these sarcomas. In addition, alternative fusion partners have been identified, including MAML3 and ZC3H7B.

BCOR-rearranged sarcoma usually occurs in bone or soft tissue of predominantly male children with a median age in the second decade of life.

There are considerable overlapping morphological and immunohistochemical features with classical Ewing sarcoma, other subtypes of small round cell tumors, as well as lymphomas and carcinomas. Therefore, the evaluation of the BCOR rearrangement status by FISH may be of diagnostic relevance.

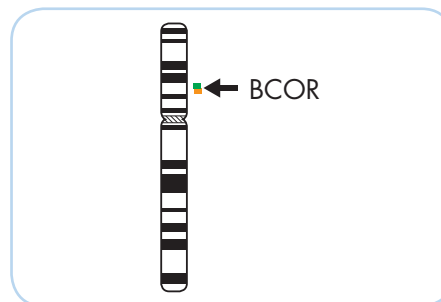
## References

- Antonescu CR, et al. (ed.) (2020) WHO Classification of Tumours Soft Tissue and Bone Tumours (5th Edition).
- Pierron G, et al. (2012) Nat Genet 44: 461-6.
- Renzi S, et al. (2019) J Cell Physiol 234: 7999-8007.
- Sirisena UDN, et al. (2021) Skeletal Radiol [Epub ahead of print].
- Specht K, et al. (2016) Am J Surg Pathol 40: 433-42.

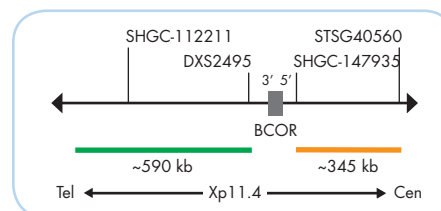
## Probe Description

The ZytoLight® SPEC BCOR Dual Color Break Apart Probe is composed of:

- ZyGreen (excitation 503 nm/emission 528 nm) labeled polynucleotides (~10 ng/µl), which target sequences mapping in Xp11.4\*\* (chrX:39,262,996-39,850,787) distal to the BCOR breakpoint region.
- ZyOrange (excitation 547 nm/emission 572 nm) labeled polynucleotides (~4.5 ng/µl), which target sequences mapping in Xp11.4\*\* (chrX:39,998,508-40,345,270) proximal to the BCOR breakpoint region.
- Formamide based hybridization buffer



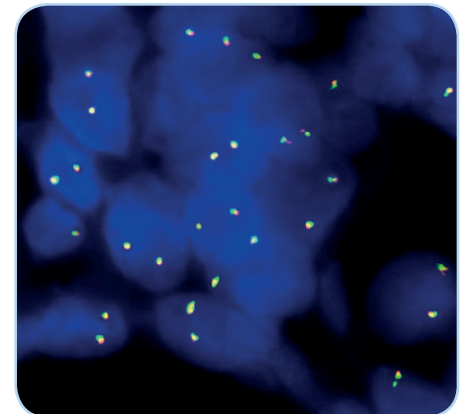
Ideogram of chromosome X indicating the hybridization locations.



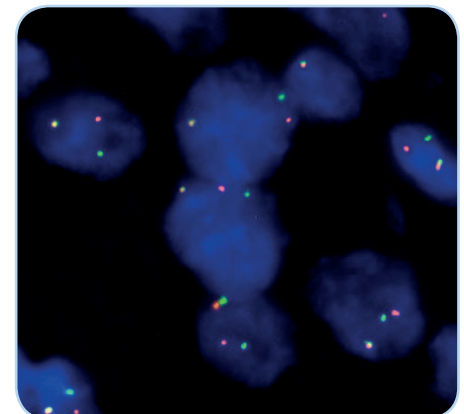
SPEC BCOR Probe map (not to scale).

## Results

In a female interphase nucleus lacking a translocation involving the Xp11.4 band, two orange/green fusion signals are expected representing two normal (non-rearranged) Xp11.4 loci. In a normal male interphase nucleus one orange/green fusion signal is expected representing one normal (non-rearranged) Xp11.4 locus. One separate green and separate orange signal indicate one Xp11.4 locus affected by a translocation or inversion.



SPEC BCOR Dual Color Break Apart Probe hybridized to normal interphase cells as indicated by two orange/green fusion signals per nucleus.



Sarcoma tissue section with translocation affecting the BCOR gene as indicated by one non-rearranged orange/green fusion signal, one orange and one separate green signal indicating the translocation.

Prod. No.	Product	Label	Tests* (Volume)
Z-2310-50	ZytoLight SPEC BCOR Dual Color Break Apart Probes CE IVD	●/●	5 (50 µl)
<b>Related Products</b>			
Z-2028-5	ZytoLight FISH-Tissue Implementation Kit CE IVD		5
Incl. Heat Pretreatment Solution Citric, 150 ml; Pepsin Solution, 1ml; Wash Buffer SSC, 210 ml; 25x Wash Buffer A, 50 ml; DAPI/DuraTect-Solution, 0.2 ml			

\* Using 10 µl probe solution per test. IVD labeled products are only available in certain countries. All other countries research use only! Please contact your local dealer for more information.

\*\*According to Human Genome Assembly GRCh37/hg19