Patient with HLH

- male
- albinism

5 ml EDTA blood

- SAP, XIAP expression

Perforin expression

- deficient

Resting NK degranulation

- normal
- abnormal

Activated NK degranulation

CTL degranulation

XLP1/2 deficient

Confirmatory tests:
- 2B4 stimulated NK cytotoxicity (XLP1)
- AIMD (XLP2)

FHL2

FHL3-5 likely

FHL unlikely

CHS/GS2

FHL likely

Confirmatory tests:
- NK cytotoxicity
- CTL cytotoxicity