

## Supplementary Figure Legends

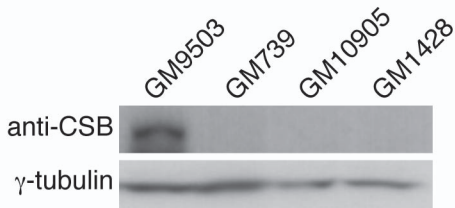
**Figure S1.** Western analysis of CSB expression in normal and CS primary fibroblasts. Immunoblotting was carried out with anti-CSB or anti- $\gamma$ -tubulin antibody. The latter was used as a loading control.

**Figure S2.** (A) Analysis of hTERT expression in GM10905 cells. One thousand cells of hTERT-immortalized GM10905 cells were used to measure telomerase activity. TSR8 was used as a positive control whereas 3-[(3-cholamidopropyl)-dimethylammonio]-1-propanesulfonate buffer (CHAPS) was used as a negative control. (B) Analysis of metaphase chromosomes from hTERT-GM10905. Chromosomes were stained with DAPI and false colored in red. Telomeric DNA was detected by FISH using a FITC-conjugated (CCCTAA)<sub>3</sub>-containing PNA probe (green).

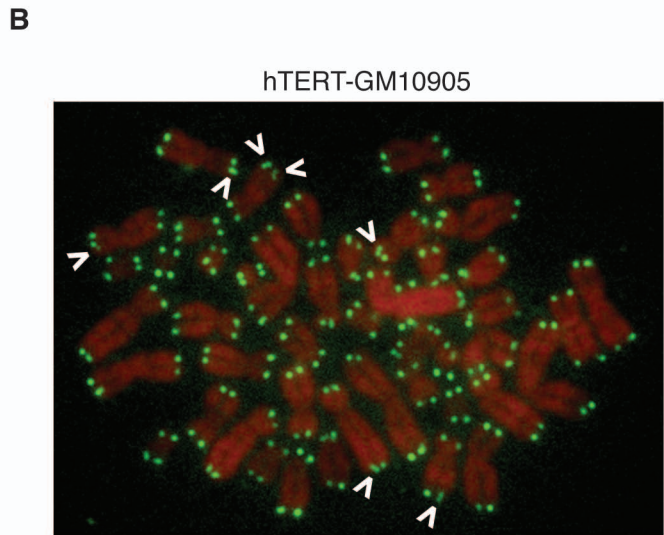
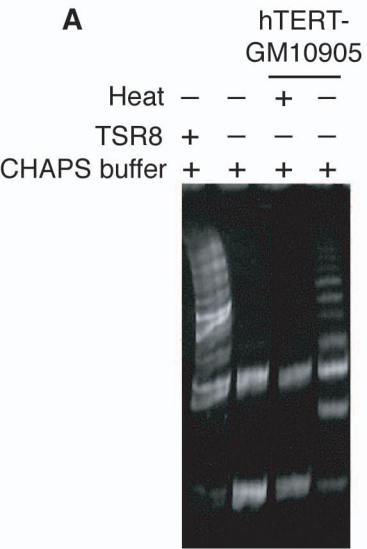
**Figure S3.** Analysis of metaphase chromosomes from hTERT-GM10905 stably expressing either the vector alone or wild type CSB. Chromosomes were stained with DAPI and false colored in red. Telomeric DNA was detected by FISH using a FITC-conjugated (CCCTAA)<sub>3</sub>-containing PNA probe (green). Open arrows represent telomere doublets.

**Figure S4.** Analysis of metaphase chromosomes from HeLaI.2.11 stably expressing either the vector alone or pRS-shCSB. Chromosomes were stained with DAPI and false colored in red. Telomeric DNA was detected by FISH using a FITC-conjugated (CCCTAA)<sub>3</sub>-containing PNA probe (green). Open arrows represent telomere doublets.

# Supplementary Figure S1 Batenburg et al.

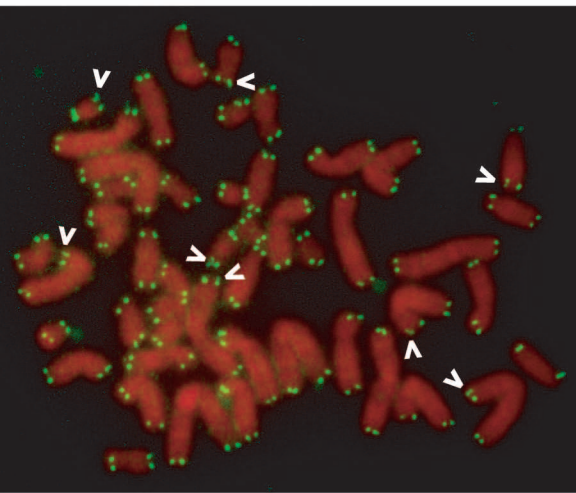


# Supplementary Figure S2 Batenburg et al.

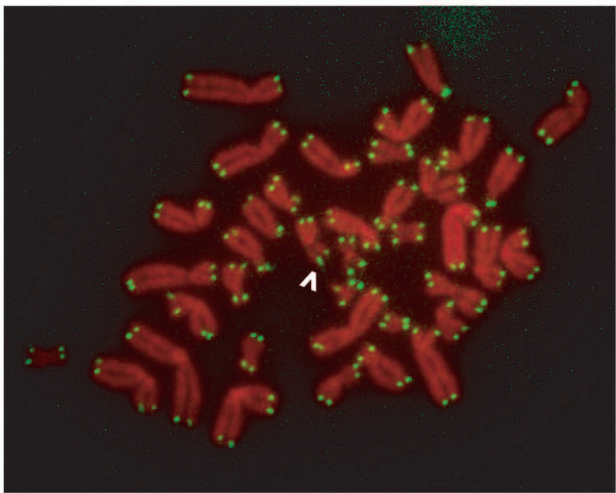


Supplementary Figure S3 Batenburg et al.

hTERT-GM10905-vector

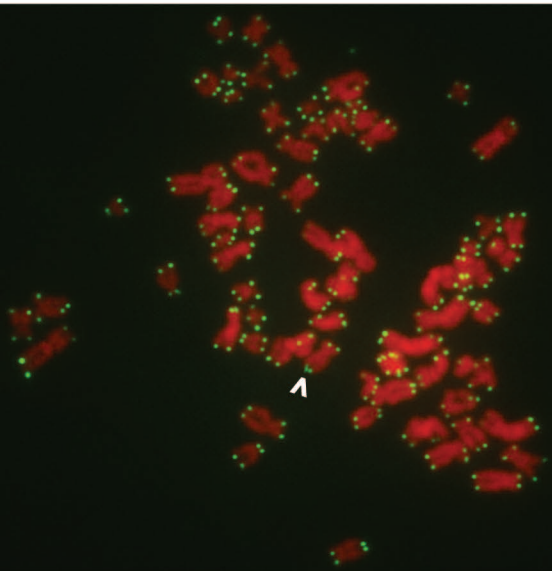


hTERT-GM10905-CSB

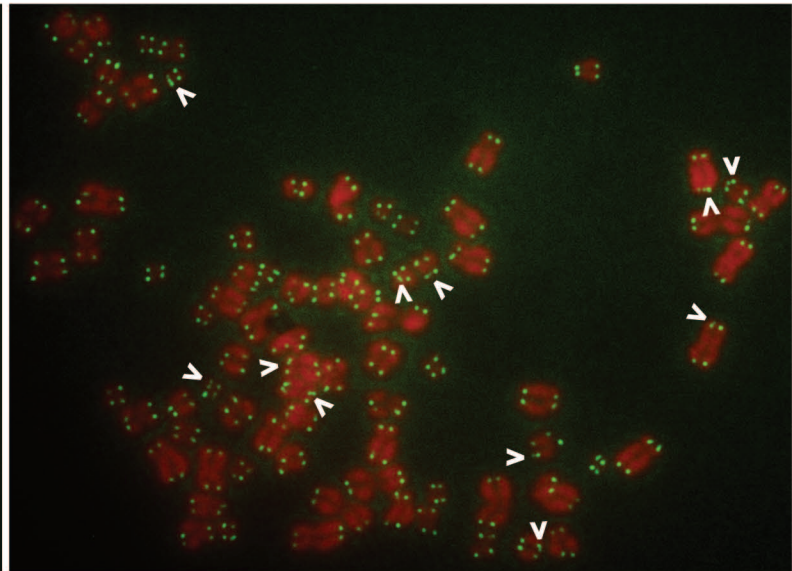


Supplementary Figure S4 Batenburg et al.

vector



shCSB



HeLa1.2.11

Supplementary Table S1. Summary of normal and CS primary fibroblast cell lines used.

| Cell Line | Age at Biopsy | CSB Allele 1 | CSB Allele 2 | Source |
|-----------|---------------|--------------|--------------|--------|
| GM38      | 9 YR          | Normal       | Normal       | NIGMS  |
| GM9503    | 10 YR         | Normal       | Normal       | NIGMS  |
| GM8399    | 19 YR         | Normal       | Normal       | NIGMS  |
| GM10901   | 42 YR         | R735X        | Normal       | NIGMS  |
| GM10905   | 10 YR         | R735X        | R735X        | NIGMS  |
| GM739     | 3 YR          | K377X        | R857X        | NIGMS  |
| GM1428    | 8 YR          | N/A          | N/A          | NIGMS  |