

Clinical laboratory values (blood and fibroblasts)

| | nmol 1/2 cystine/mg protein |
|----------------------|------------------------------------|
| c.530 A>G WBC | 1.31 |
| c.530A>G fibroblasts | 3.4 |

c.530 A>G ciPTEC and ciPODO measurements

| | Solution | Total Volume NEM+SSA (μl) | Protein μg/mL | Cys μM | Cys nmol | Cys nmol/mg protein | 1/2 cystine nmol/mg protein | average | stdev |
|-----------------|-----------------|----------------------------------|----------------------|---------------|-----------------|----------------------------|------------------------------------|----------------|--------------|
| c.530 A>G PTECs | 5mM NEM+12%SSA | 100+50 | 563.8 | 4.02 | 4018.304 448 | 7.13 | 14.25 | 13.39 | 0.69 |
| c.530 A>G PTECs | 5mM NEM+12%SSA | 100+50 | 806.1 | 5.04 | 5035.517 598 | 6.25 | 12.49 | | |
| c.530 A>G PTECs | 5mM NEM+12%SSA | 100+50 | 752.4 | 5.13 | 5129.770 467 | 6.82 | 13.64 | | |
| c.530 A>G PTECs | 5mM NEM+12%SSA | 100+50 | 776.4 | 4.90 | 4903.241 845 | 6.32 | 12.63 | | |
| c.530 A>G PTECs | 5mM NEM+12%SSA | 100+50 | 811.1 | 5.49 | 5486.540 248 | 6.76 | 13.53 | | |
| c.530 A>G PTECs | 5mM NEM+12%SSA | 100+50 | 728.9 | 5.04 | 5035.963 029 | 6.91 | 13.82 | | |
| c.530 A>G PODOs | 5mM NEM+12%SSA | 100+50 | 706.3 | 2.73 | 2733.833 538 | 3.87 | 7.74 | 8.59 | 0.62 |
| c.530 A>G PODOs | 5mM NEM+12%SSA | 100+50 | 625.8 | 2.72 | 2719.969 076 | 4.35 | 8.69 | | |
| c.530 A>G PODOs | 5mM NEM+12%SSA | 100+50 | 659.9 | 3.10 | 3099.073 063 | 4.70 | 9.39 | | |
| c.530 A>G PODOs | 5mM NEM+12%SSA | 100+50 | 766.6 | 3.46 | 3462.062 907 | 4.52 | 9.03 | | |
| c.530 A>G PODOs | 5mM NEM+12%SSA | 100+50 | 756.2 | 3.03 | 3026.392 672 | 4.00 | 8.00 | | |
| c.530 A>G PODOs | 5mM NEM+12%SSA | 100+50 | 707.3 | 3.07 | 3072.633 352 | 4.34 | 8.69 | | |

Control values

| | Solution | Total Volume NEM+SSA (μl) | Protein μg/mL | Cys μM | Cys nmol | Cys nmol/mg protein | 1/2 cystine nmol/mg protein | average | stdev |
|---------------|----------------|------------------------------|---------------|-----------|-----------------|------------------------|--------------------------------|---------|-------|
| Control PTECs | 5mM NEM+12%SSA | 100+50 | 797.1 | 1.15 | 1152.108 162 | 1.45 | 2.89 | 2.04 | 0.68 |
| Control PTECs | 5mM NEM+12%SSA | 100+50 | 751.8 | 0.73 | 725.3667 974 | 0.96 | 1.93 | | |
| Control PTECs | 5mM NEM+12%SSA | 100+50 | 778.1 | 0.81 | 812.0816 146 | 1.04 | 2.09 | | |
| Control PTECs | 5mM NEM+12%SSA | 100+50 | 797.1 | 0.49 | 494.4965 49 | 0.62 | 1.24 | | |
| Control PODOs | 5mM NEM+12%SSA | 200+100 | 297.6 | 0.63 | 626.5411 046 | 2.11 | 4.21 | 5.01 | 1.65 |
| Control PODOs | 5mM NEM+12%SSA | 200+100 | 212.2 | 0.73 | 732.9975 204 | 3.45 | 6.91 | | |
| Control PODOs | 5mM NEM+12%SSA | 200+100 | 319.5 | 0.62 | 623.6802 777 | 1.95 | 3.90 | | |