Frataxin levels: Regression with GAA1 (nonlinear), GAA2, Age of Onset, Sex

| Cohort | TQ, FXN-E | | TQ, FXN-M | | LF, FXN-M | |
| --- | --- | --- | --- | --- | --- | --- |
| Variable | Estimate ± SE | p-value | Estimate ± SE | p-value | Estimate ± SE | p-value |
| Model | 0.32 (R²) | <0.0001 | 0.28 (R²) | <0.0001 | 0.39 (R²) | <0.0001 |
| Sex | 0.058 ± 0.035 | 0.1007 | -0.033 ± 0.034 | 0.3293 | 0.019 ± 0.028 | 0.5140 |
| AOO | *0.009 ± 0.004* | *0.0120* | *0.012 ± 0.004* | *0.0006* | *0.012 ± 0.003* | *<0.0001* |
| GAA1 (nonlinear) |  | 0.0030 |  | <0.0001 |  | 0.0068 |
| GAA2 | -0.012 ± 0.008 | 0.1551 | -0.020 ± 0.008 | 0.0123 | -0.014 ± 0.007 | 0.0649 |

Frataxin levels: Regression with GAA, Age, Sex, Identical

| Cohort | TQ, FXN-E | | TQ, FXN-M | | LF, FXN-M | |
| --- | --- | --- | --- | --- | --- | --- |
| Variable | Estimate ± SE | p-value | Estimate ± SE | p-value | Estimate ± SE | p-value |
| Model | 0.29 (R²) | <0.0001 | 0.22 (R²) | <0.0001 | 0.38 (R²) | <0.0001 |
| Sex | 0.048 ± 0.036 | 0.1830 | -0.045 ± 0.035 | 0.2001 | 0.009 ± 0.029 | 0.7449 |
| AOO | 0.004 ± 0.003 | 0.2280 | 0.006 ± 0.003 | 0.0632 | 0.010 ± 0.002 | <0.0001 |
| GAA1 | -0.059 ± 0.011 | <0.0001 | -0.036 ± 0.011 | 0.0018 | -0.056 ± 0.009 | <0.0001 |
| GAA2 | -0.013 ± 0.008 | 0.1143 | -0.022 ± 0.008 | 0.0092 | -0.017 ± 0.007 | 0.0193 |

Frataxin levels: Regression with GAA, Age, Sex, Identical

| Cohort | TQ, FXN-E | | TQ, FXN-M | | LF, FXN-M | |
| --- | --- | --- | --- | --- | --- | --- |
| Variable | Estimate ± SE | p-value | Estimate ± SE | p-value | Estimate ± SE | p-value |
| Model | 0.28 (R²) | <0.0001 | 0.22 (R²) | <0.0001 | 0.38 (R²) | <0.0001 |
| AOO | 0.0039 ± 0.0034 | 0.2552 | 0.0072 ± 0.0034 | 0.0323 | 0.0092 ± 0.0023 | <0.0001 |
| GAA1 | -0.0596 ± 0.0131 | <0.0001 | -0.0256 ± 0.0129 | 0.0480 | -0.0595 ± 0.0106 | <0.0001 |
| GAA2 | -0.0147 ± 0.0106 | 0.1666 | -0.0304 ± 0.0104 | 0.0038 | -0.0139 ± 0.0084 | 0.0998 |
| Identical | -0.0071 ± 0.0552 | 0.8976 | -0.0777 ± 0.0543 | 0.1539 | 0.0277 ± 0.0443 | 0.5326 |

Frataxin levels: Regression with GAA, Age, Sex, Identical

| Cohort | TQ, FXN-E | | TQ, FXN-M | | LF, FXN-M | |
| --- | --- | --- | --- | --- | --- | --- |
| Variable | Estimate ± SE | p-value | Estimate ± SE | p-value | Estimate ± SE | p-value |
| Model | 0.29 (R²) | <0.0001 | 0.23 (R²) | <0.0001 | 0.38 (R²) | <0.0001 |
| Sex | 0.0478 ± 0.0357 | 0.1825 | -0.0438 ± 0.0351 | 0.2131 | 0.0103 ± 0.0286 | 0.7185 |
| Age | 0.0010 ± 0.0025 | 0.6873 | -0.0031 ± 0.0024 | 0.1959 | 0.0023 ± 0.0016 | 0.1354 |
| AOO | 0.0030 ± 0.0045 | 0.5010 | 0.0107 ± 0.0044 | 0.0158 | 0.0068 ± 0.0029 | 0.0196 |
| GAA1 | -0.0583 ± 0.0131 | <0.0001 | -0.0267 ± 0.0129 | 0.0395 | -0.0578 ± 0.0107 | <0.0001 |
| GAA2 | -0.0140 ± 0.0106 | 0.1891 | -0.0320 ± 0.0104 | 0.0024 | -0.0142 ± 0.0084 | 0.0939 |
| Identical | -0.0072 ± 0.0556 | 0.8974 | -0.0829 ± 0.0545 | 0.1296 | 0.0243 ± 0.0445 | 0.5846 |