

## **Supplemental Information**

### **Alleviating GAA repeat induced transcriptional silencing of the Friedreich's ataxia gene during somatic cell reprogramming**

Urszula Polak<sup>1</sup>, Yanjie Li<sup>2</sup>, Jill Sergesketter Butler<sup>2\*</sup>, and Marek Napierala<sup>2,3\*</sup>

1. Supplemental Table 1
2. Supplemental Figure 1
3. Supplemental Figure 2
4. Supplemental Figure 3

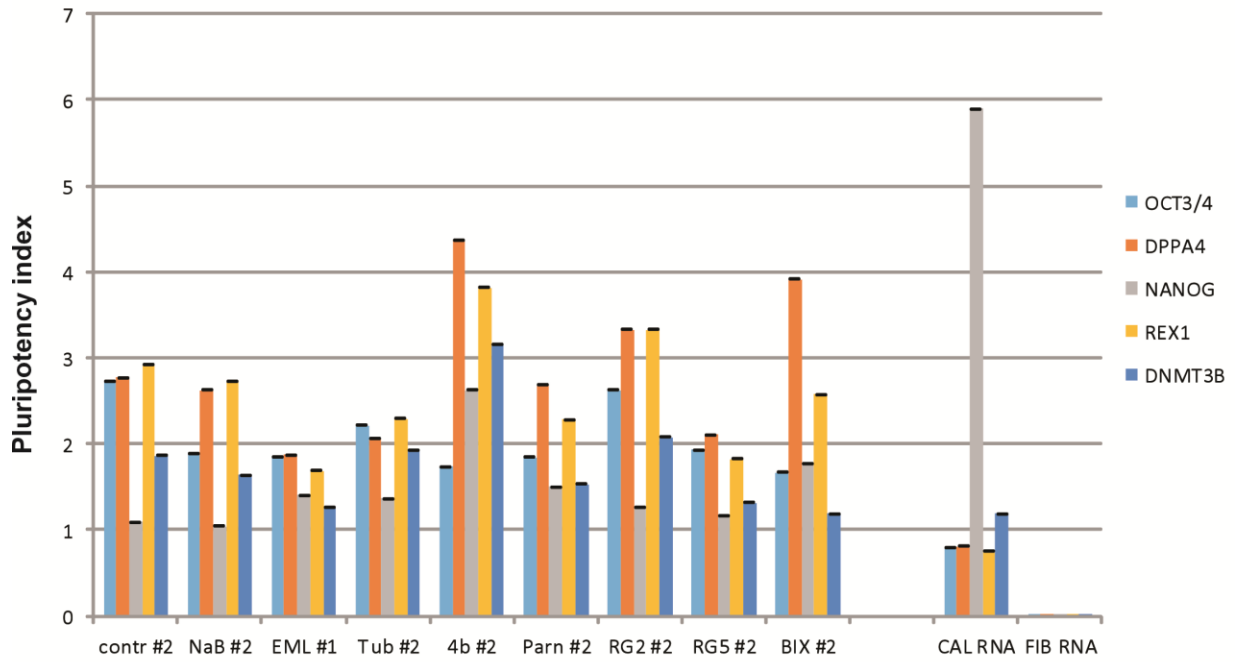
Supplemental Table 1. Primers used in this study.

Primer	Forward 5 → 3	Reverse 5 → 3
<i>To determine number of GAA repeats</i>		
FXN_short	GGCTTGAACTTCCCACACGTGTT	AGGACCATCATGGCCCACTT
<i>Quantitative RT-PCR for FXN mRNA</i>		
<i>Frataxin</i>	CAGAGGAAACGCTGGACTCT	AGCCAGATTTGCTTGTTTGG
<i>GAPDH</i>	GAAGGTGAAGGTCGGAGTC	GAAGATGGTGATGGGATTTC
<i>β-actin</i>	CTTCCTGGGCATGGAGTC	ATCTTGATCTTCATTGTGCTG
<i>L19</i>	CTGAAGGTGAAGGGGAATGTG	GGATAAAGTCTTGATGATCTC
<i>Quantitative RT-PCR for retroviral transgene silencing</i>		
pMXs-F	GTGGTGGTACGGGAAATCAC	
pMX-Oct3/4		TAGCCAGGTTTCGAGAATCCA
pMX-Klf4		GGGAAGTCGCTTCATGTGAG
pMX-Sox2		GGTTCTCCTGGGCCATCTTA
pMXs-Myc		AGCAGCTCGAATTTCTTCCA
<i>FXN CHIP primers</i>		
Promoter	CCCCACATACCCAAGTCTG	GCCCGCCGCTTCTAAAATTC
Upstream	GAAACCCAAAGAATGGCTGTG	TTCCCTCCTCGTGAAACACC
Downstream	CTGGAAAAATAGGCAAGTGTGG	CAGGGGTGGAAGCCCAATAC

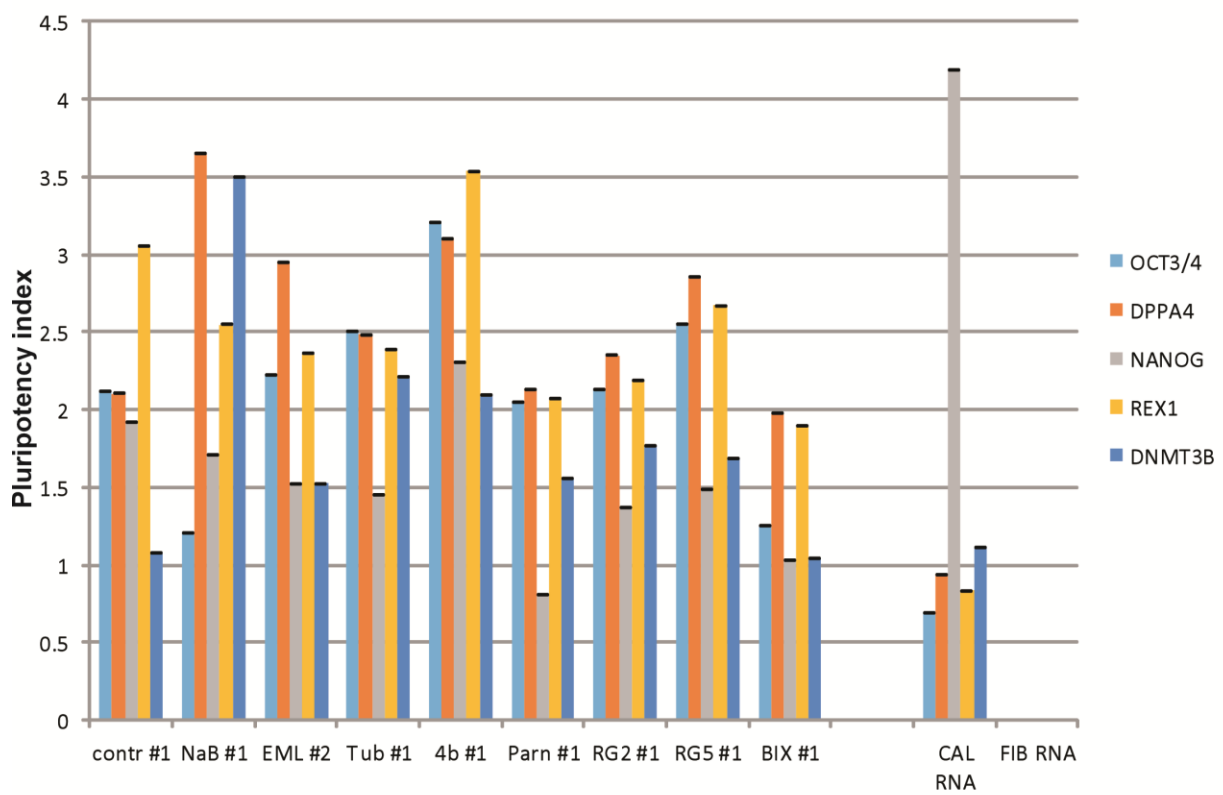
*Supplemental Figure 1. Determination of pluripotency marker expression using the PluriPCR™ kit for iPSC lines generated in this study.* The expression of five genes: Oct-3/4 (light blue bars), Nanog (grey bars), DNMT3b (dark blue bars), Dppa4 (orange bars), and Rex1 (yellow bars) that are strongly indicative of pluripotency was quantified. Results were evaluated using the MTI-GlobalStem provided template. Each group represents an example of the analysis of each individual iPSC clone reprogrammed in the presence of DMSO (contr), NaB, EML, Tub, 4b, Parn, RG and BIX (abbreviations designated in Table 1). “CAL RNA” and “FIB RNA” represent manufacturer supplied positive and negative control RNAs isolated from embryonic stem cells and fibroblast cells, respectively. Panels A and B present two independent experiments on separate iPSC clones.

# Supplemental Figure 1

A

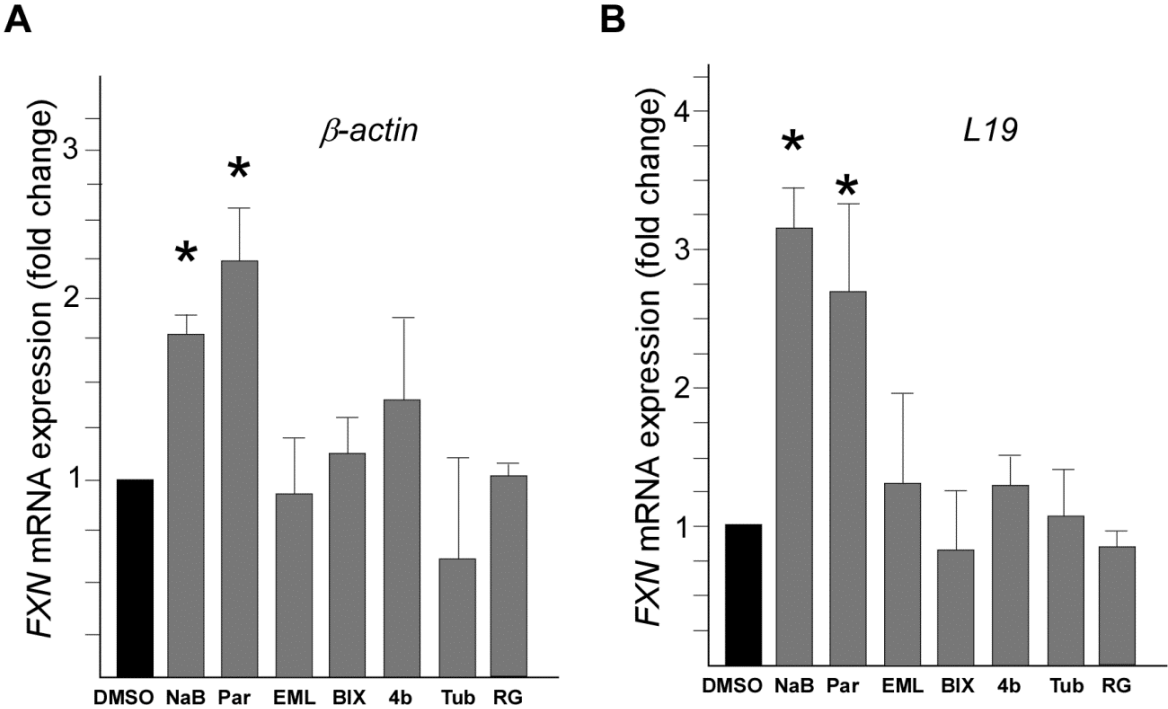


B



Supplemental Figure 2. qRT-PCR analyses of FXN mRNA expression in FRDA iPSCs reprogrammed in the presence of epigenetic inhibitors. FXN transcript levels were normalized relative to  $\beta$ -actin mRNA (A) and L19 RNA (B). Primers used in the analyses are given in Supplementary Table 1.

### Supplemental Figure 2



Supplemental Figure 3. Expression of FXN mRNA in control iPSCs (GM08399) harboring short GAA repeats. No differences in FXN transcript levels were detected between passages 2, 9, 14 and 26.

### Supplemental Figure 3

