F806 Analysis of the *FBN1* intragenic microsatellite polymorphisms in Koreans

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Fibrillin-1 (FBN1) is a large glycoprotein (320 kDa) ubiquitously distributed in connective tissue. The gene encoding FBN1 is located at chromosome 15q21.1 and is relatively large (110 kb) and highly fragmented with 65 exons. Mutations in the *FBN1* gene have been found to cause Marfan syndrome (MFS) that is an autosomal dominant disorder affecting the cardiovascular, skeletal, and ocular systems. We identified the four intragenic microsatellites of the *FBN1* intron 1, 5, 28, and 43 (mts-1, 2, 3, and 4) for genetic markers in Koreans. The number of distinct marker alleles for intragenic microsatellites in *FBN1* intron 1, 5, 43 (CA repeat), and 28 (TAAAA repeat) showed 8, 12, 9, and 2, respectively. The most frequent alleles for the mts-1, 2, 3, and 4 have frequencies of 0.32, 0.20, 0.53, and 0.60, respectively. The observed heterozygosity were 0.78, 0.87, 0.50, and 0.61, respectively and there were differences from that of Cauccsians, concerning allele frequency, number of alleles, and heterozygosity.

F807 **Requiem Expression is Regulated at the Translation Level by a Cis-acting Element in Its 3' Untranslated Region and a Cellular 55 KDa Trans-acting Factor**

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Requiem, originally cloned in IL-3 derived murine myeloid cell line FDCP-1, is a novel zinc finger protein essential for apoptosis in myeloid cells. On the other hand, we identified a clone G8 that encodes a part of 3' untranslated region (UTR) of requiem as a putative β-globin gene switching factor. Practically, we confirmed the overexpression of G8 RNA induced human β-globin expression in K562 cell line in which fetal-type γ-globin was normally expresse, even though expression of endogenous requiem at transcription level was not modulated during *in vitro* differentiation of erythroid cells. From the previous data that an antisense 3UTR of requiem rescues FDCP-1 cell line from apoptosis, and our data that overexpression of G8 mediates β-globin switching, we speculate that translational control may be important for the regulation of requiem gene expression. Here, we present evidence that the 3'UTR of requiem, along with its trans-acting factor, has an ability to repress translation. The translation of enhanced green fluorescence protein (EGFP) reporter linked to full length 3'UTR was severely repressed, but the transcriptional level of EGFP was similar to the control vector not containing G8 region. In addition, the crucial region for this inhibition was identified by assaying the translational level of EGFP linked to serially deleted mutants of G8. To verify the presence of factor(s) binding to specific G8 region, EMSA and UV crosslinking assay were performed. A cellular factor, about 50 KDa, is identified and its 66 bp core binding site is coincide with the G8 region that mostly inhibits the translation of reporter. RNA secondary structure was also crucial for the interaction between trans-acting factor and G8 region. Taken together, this data suggests that specific protein binding to the sequences within requiem 3'UTR regulates requiem synthesis, eventually causing apoptosis in myeloid cells or β-globin switching in erythroid cells.