ADDENDUM

Chapter I:

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CHAPTER II:

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There were 29 patients, all independent of blood transfusions in Group C. Of these patients 5 harbored co inherited alpha thalassemia with a 3.7 Kb deletion and 1 had a 4.2 Kb deletional alpha thalassemia, and this brings the incidence of the alpha thalassemia to 6 in this group. So 23 patients out of 29 i.e., 79.3% of these patients were not having a coinherited alpha thalassemia in this non transfused group. Again 6 patients in this group had the +/- status of the XmnI polymorphism, 23 had the +/- status and none had the -/- status which again brings the incidence of +/- status of the XmnI polymorphism to 20.7% and the percentage of patients having the +/- status was 79.3%. The patients not harboring any deletional alpha thalassemia mutation or having the +/- XmnI status might have inherited non deletional alpha thalassemia to decrease the severity of the disease or some other factors led to the their milder clinical course. Factors or genetic loci inside or outside the beta globin gene cluster modulating the levels of fetal hemoglobin might also be responsible for the lessened severity of the disease in these patients.
It can be seen from the Table 2.4 that the patients have been clearly divided into three distinct groups A, B, and C based on their transfusion dependency and the respective mutations and the number of patients having the same set of mutations in an individual group have been clearly shown in the table 2.4.
Indeed, the probability of chromosome 6 being different in the two siblings cannot be totally excluded and other variable number of tandem repeats that segregate between the two parents and fingerprinting using other such VNTR markers might be able to resolve this discordant inheritance pattern in the two brothers.

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