Correspondence

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Did the Hb G San José gene arise in Sicily?

SIR,

We have studied 969 school children from Grammichele, a small town of 13 000 inhabitants 50 km from Catania. These children were screened for abnormal haemoglobins using a Coulter Counter and cellulose acetate electrophoresis. Structural analysis of haemoglobin was performed when required. The results are summarised in the table.

We found seven children from seven different apparently unrelated families with Hb G San José trait and another 19 subjects when their families were studied. These 26 patients showed no clinical or haematological anomalies apart from the Hb G San José concentration levels, which varied from 33.4% to 36.9%.

Grammichele was rebuilt in its beautiful hexagonal architectural style after a devastating earthquake in 1693 which killed nearly all its inhabitants.¹ In order to populate the new town, Carlo II of Austria issued a decree giving asylum to criminals and other

fugitives.² Since then there has been almost no immigration and the frequency of consanguineous marriages in the area has been quite high, varying from 0.39% (1780 to 1785) to 9.11% (1918 to 1924) of the total marriages; the frequency now is about 4.73%.³ We feel that this cluster of Hb G San José in a small town is very surprising and constitutes an example of 'founder effect'.

It must be remembered that eight of the other nine families with this haemoglobinopathy described so far are of Sicilian or Calabrian origin.^{4,5} Calabria is the southern tip of the Italian peninsula, separated from Sicily only by the narrow Straits of Messina. This would suggest that the gene for Hb G San José originated in our region.

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TABLE Summary of survey results on 969 subjects examined.

	No	%
β thalassaemia trait	56	5.77
Hb G San José	7	0.72
Hb S	7	0.72
Hb J Oxford	1	0.10

References

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- ³ Moroni A, Menozzi P. La consanguineità umana in Sicilia. *Ateneo Parmese* 1972;8:suppl 1.
- ⁴ Schilirò G, Li Volti S, Musumeci S, et al. Sicily: a cluster of Hb G San José. *Hemoglobin* 1981;5:725-30.
- ⁵ Marinucci M, Schilirò G, Russo G, Tentori L. Hemoglobinopathies in Italy. *Hemoglobin* 1982;6:247-55.