Correspondence

Clinical features of homozygous α2(I) collagen deficient osteogenesis imperfecta

SIR,

You recently published our reply to Mr Gage's comments (J Med Genet 1985;22:319–20) that our patient with severe Sillence type III osteogenesis imperfecta and a homozygous four base deletion at the 3' end of the collagen α2(I) gene (J Med Genet 1984;21:257–62) could have had dentinogenesis imperfecta despite our clinical comment to the contrary. We then published pictures which supported our statement rather than his, but accepted that this did not place the matter beyond dispute.

We have now been fortunate in acquiring a tooth from the affected patient and in collaboration with the Eastman Dental Hospital have studied the morphology of the dentine in the deciduous incisor. These studies confirm that our patient has a normal pattern of dentine when compared with a deciduous incisor with typical dentinogenesis imperfecta and a normal control (figure).

F M Pope*, A C Nicolls*, G Osse†, and K W Lee‡
*Dermatology Research Group, Clinical Research Centre, Harrow;
†Kinderklinik und Poliklinik, Göttingen, Germany; and
‡Eastman Dental Hospital, Institute of Dental Surgery, London.

FIGURE (Top) Section from tooth with typical dentinogenesis imperfecta showing a normal zone of mantle dentine at the periphery with grossly abnormal circumpulpal dentine compared with (middle) the pattern from the homozygous α2 deficient OI patient and (bottom) a normal deciduous incisor. All sections were stained with picrothionin.