“...normal pulp cavities exclude dentinogenesis imperfecta”.

In the dentine abnormalities that may be associated with osteogenesis imperfecta, the pulp chambers and root canals are either progressively obliterated by the continuous deposition of highly disorganised dentine, or the pulp chambers are larger than normal by a failure of deposition. It is not possible to ascertain whether or not the pulp chambers are in fact enlarged, or to measure the amount of obliteration without taking periapical radiographs and comparing them with age matched controls.

There is also no mention in the article about tooth crown morphology or if, in fact, the teeth are smaller with cusp tips closer together than normal. Nor is there any reference to the presence or absence of short tapering roots.

Although discoloration and opalescence are the more easily observed signs of abnormality, they are not the only criteria for the classification of dentinogenesis imperfecta.

J P GAGE
Nuffield Orthopaedic Centre, Headington, Oxford OX3 7LD.

This letter was shown to Dr Nicholls and colleagues, who reply as follows:

SIR,

We note Mr Gage's comments with interest and fully accept that the diagnosis of dentinogenesis imperfecta can be difficult, especially for non-dentists. Undoubtedly the assessment of the finer details of pulp cavity and root canal morphology is the province of dental expertise and well beyond the abilities of the casual observer. Yet most clinicians in the field rely upon tooth colour to diagnose dentinogenesis perhaps more than they ought.

Unfortunately panorthotomograms were not available from our patient and we have been unable to obtain a tooth for histological examination (which would settle matters beyond dispute). We therefore showed the clinical morphology of our patient's teeth and their appearance on skull x-rays to established dental experts interested in dentinogenesis in the London region. In their opinion neither the clinical appearance (fig 1) nor the tooth radiographs (fig 2) show any evidence of dentinogenesis imperfecta in this patient.

F M POPE AND A C NICHOLLS
Dermatology Research Group, Clinical Research Centre, Watford Road, Harrow, Middlesex HA1 3UJ.

HLA antigens in South African Afrikaners with heterozygous familial hypercholesterolaemia

SIR,

Familial hypercholesterolaemia (FH) is inherited as an autosomal dominant disorder. The prevalence of FH heterozygotes in the white Afrikaner population of South Africa is 1 in 100 or more and is the highest ever recorded.1 We have investigated the distribution of HLA antigens in 82 unrelated

FIG 1. Clinical appearance of patient showing apparently normal teeth morphology and colour.

FIG 2. A, P, and lateral views of patient's dentition enlarged from original skull x-ray.
Afrikaner FH heterozygotes living in the southern Transvaal who attended the Lipid Disorders Clinic of the Johannesburg Hospital and compared it with that in 278 healthy unrelated white persons residing in the Johannesburg area.

The following HLA-A and B antigens were determined by means of standard microlymphocytotoxicity tests, modified by incubation at 37°C throughout the procedure, and using trypan blue as the indicator dye: A1, A2, A3, A9, A10, A11, A28, A29, Aw30, Aw31, Aw32, Aw33, Aw43, B5, B7, B8, B12, B13, B14, B15, Bw16, B17, B18, Bw21, Bw22, B27, Bw35, B37, B40, Bw41, and Bw42. The divergence of the frequency of any antigen of the patient group and the control group was determined by using standard statistical analysis.

No antigen occurred more frequently among the FH heterozygotes. HLA-Aw32 and HLA-Bw22 occurred less frequently (table) but in view of the small percentages of patients who possessed these antigens this finding is probably not important. Furthermore, these differences are no longer apparent when the number of antigens tested is taken into account. We conclude that there is no significant association or link between the HLA antigens tested and heterozygous FH.

S G BAKER, A RABSON*, R SHIRES, B I JOFFE, AND H C SEFTEL
Carbohydrate and Lipid Metabolism Research Group, Department of Medicine, University of the Witwatersrand Medical School, and
*Department of Immunology, School of Pathology, South African Institute of Medical Research, Johannesburg, South Africa.

References

Prenatal screening for Down syndrome

SIR,

The proposal by Dr Cuckle and colleagues (Lancet, 28 April 1984, p 926)* that 21% of Down syndrome (DS) fetuses could be diagnosed if women whose midtrimester serum alphafetoprotein (MSAFP) levels were ≤0.5 MOMs (multiples of the median for gestational age) were offered amniocentesis is attractive. A simple prenatal screening test for DS is needed urgently, but the implications of the proposal should be considered carefully.

I have examined what would have happened had such a scheme been in operation during the five completed years of MSAFP screening for neural tube defects (NTD) at Guy’s Hospital, London, all women having been delivered. There were a total of 9788 deliveries, and this population contained 12 DS babies (one in 816); 6541 (67%) women had their MSAFP tested between the 15th and the 20th gestational week (assayed in the Department of Clinical Chemistry), and 472 (7%) were ≤0.5 MOMs. Eight of the 12 mothers with DS fetuses were screened (table 1), one of whom (13%) was below the cut-off point. If a screening policy based on a combination of maternal age and a variable cut-off were adopted, as further suggested by Cuckle et al, then another DS (0.79 MOMs, maternal age 36) were diagnosed.


Letter reprinted from The Lancet with permission of the author and publisher.