Coeliac disease in identical twin infants

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Summary

A pair of monozygous twins had coeliac disease in infancy, and all of their family carried the HLA-B8 antigen. Both genetic and environmental factors were involved in the aetiology.

Introduction

Coeliac disease (CD) is familial, and most cases carry the HLA-Dw3 allele; 10% of subjects from the families affected by the disease have jejunal villous atrophy (Dennis and Stokes, 1978). Identical twins with CD whose genotype was homozygous HLA1-B8 are now reported.

Case reports

Male twins, aged 19 years

Both boys vomited during weaning at the age of 7 months. They were admitted 10 months later because of weight loss and recurring vomiting, and their faecal-fat excretion was 44% and 48%. Both children recovered when given a gluten-free diet. At the age of 11 years, their bone age was 10 years. They then returned to a normal diet, although continuing to have gluten-free bread. At 19 years old, one twin complained of tiredness, and the other was symptomless. Their haemoglobin was 11.2 and 10.4 g/dl respectively, and they had diminished xylose excretion. Their serum IgA was 0.6 g/l and 0.8 g/l (normal 1.0-4.3 g/l). Both twins were shown by jejunal biopsy to have subtotal villous atrophy while on a normal diet. Their blood groups (ABO, RL., MNS., S., P. Lu., K., Le., Ky., Jk.) and HLA tissue type were identical (Table 1).

Their sister, aged 21 years, was fit and not anaemic, and her serum IgA was 1.2 g/l.

TABLE 1. HLA-B antigens of family with coeliac disease

	Age	Tissue-type
Father	50	HLA3, A11, B8
Mother	52	HLA1, B8, BW15
Daughter	21	HLA1, A3, B8, BW15
Twin I	19	HLA1, A3, B8
Twin II	19	HLA1, A3, B8

} Coeliac disease

Their mother, aged 52 years, had no anaemia and was healthy, but her serum IgA was 0.8 g/l. At the age of 15 years she was ill for 6 weeks with a fever, erythema nodosum and arthralgia.

Their father, aged 50 years, had no illness, and his Hb, xylose absorption and serum IgA were normal. His chest X-ray showed small calcified lesions throughout both lungs, and his Tine test was strongly positive. There was a monoclonal IgG paraprotein band in his serum. All members of the family had the autoimmune tissue-type HLA-B8 (Table 1).

Discussion

One or both of 11 pairs of twins have been reported as having CD (Hoffman, Wollaeger and Greenberg, 1966; Walker-Smith, 1973; Lewkowicz, Gairdner and Doe, 1976; Penna *et al.*, 1979). Four of these were discordant, and 7 were concordant for the disease. Six of the pairs were identical twins, and the remaining 5 may also have been monozygotic. Neither the present twins nor their mother mounted a normal IgA response to ingested antigens (Hodgson, Davies and Gent, 1976) and all had serum IgA deficiency. They were the second and third siblings in the family. The second and third children in another family also had the disease, and they were not twins. Their parents had no haematological or biochemical changes, nor did they carry the HLA-B8 allele. The mother was tissue type HLA1-B8 and the father was HLA9-B35. Both genetic and environmental factors are involved in CD. HLA-B8-linked autoimmune disease associated with defective Fc-receptor function (Lawley *et al.*, 1981), and premature weaning with gluten-containing foods (Littlewood and Crollick, 1980) are probably important in the aetiology. The mother of the present twins possibly had sarcoidosis, and this disease is sometimes associated with CD (MacGregor, 1976).

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References

- DENNIS, N.R. & STOKES, C.R. (1978) Risk of C.D. in children of patients and effect of HLA genotype. *Journal of Medical Genetics*, **15**, 20.
- HODGSON, H.J.F., DAVIES, R.J. & GENT, A.E. (1976) Atopic disorders and adult coeliac disease. *Lancet*, **i**, 115.
- HOFFMAN, H.N., WOLLAEGER, E.E. & GREENBERG, E. (1966) Discordance for coeliac disease in a monozygous twin pair. *Gastroenterology*, **51**, 36.
- LAWLEY, T.J., HALL, R.P., FAUCI, A.S., KATZ, S.I., HAMBURGER, M.I. & FRANK, M.M. (1981) Defective Fc-receptor functions associated with the HLA-B8/DRw3 haplotype. Studies in patients with dermatitis herpetiformis and normal subjects. *New England Journal of Medicine*, **304**, 185.
- LEWKONIA, R.M., GAIRDNER, D. & DOE, W.F. (1976) IgA deficiency in one of identical twins. *British Medical Journal*, **1**, 311.
- LITTLEWOOD, J. & CROLLICK, A.J. (1980) Childhood coeliac disease is disappearing. (letter) *Lancet*, **ii**, 1359.
- MACGREGOR, G.A. (1976) Gluten and lymphocytes in coeliac disease. *Lancet*, **ii**, 860.
- PENNA, F.J., MOTA, J.A.C., ROQUETE, M.L.V., DE CARVALHO, A.S.T., LEMOS, A.T.de O., BARBOSA, A.J.F., LEAO, E., FERREIRA, R.A. & CASTRO, L. De P. (1979) Coeliac disease in identical twins. *Archives of Disease in Childhood*, **53**, 395.
- WALKER-SMITH, J.A. (1973) Discordance for childhood coeliac disease in monozygous twins. *Gut*, **14**, 374.