```
bed PCVISREIMENYNIALRWTAKQKLYSRTGESVEFVCKRGYRLSSRSHTLRTTCWDGKLEFQS.....
hfr PCVISREIMENYNIALRWTAKQKLYLRTGESAEFVCKRGYRLSSRSHTLRTTCWDGKLEYPTCAKR.
HF  PCVISREIMENYNIALRWTAKQKLYSRTGESVEFVCKRGYRLSSRSHTLRTTCWDGKLEYPTCAKR.
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Figure 2 Alignment of the amino acid sequence of exon 20 in the Bedouin family, FHR1 exon 5 and HF exon 20. The two amino acid differences between FHR1 exon 5 and HF exon 20 are in the boxes. (Abbreviations: bed, Bedouin family; hfr, factor H-related protein 1; HF, Factor H)

ulonephritis rather than HUS? These unanswered questions emphasize the need for further investigation of the genotype-phenotype correlations in FH abnormalities.

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MARK R. H. BUDDLES,[1] ROSEMARY L. DONNE,[1,2] ANNA RICHARDS,[1,2] JUDITH GOODSHIP,[1] AND TIMOTHY H. J. GOODSHIP[2]

[1]*School of Biochemistry and Genetics and* [2]*School of Clinical Medical Sciences, The Medical School, University of Newcastle upon Tyne, Newcastle upon Tyne, United Kingdom*

Electronic-Database Information

The URL for data in this article is as follows:

Online Mendelian Inheritance in Man (OMIM), http://www.ncbi.nlm.nih.gov/Omim (for HUS [MIM 235400])

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Address for correspondence and reprints: Timothy H. J. Goodship, Royal Victoria Infirmary, Newcastle upon Tyne, NE1 4LP, United Kingdom. E-mail: t.h.j.goodship @ncl.ac.uk

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Reply to Buddles et al.

To the Editor:

We congratulate Buddles et al. on their thorough evaluation of the factor H gene and on the identification of a mutation that we failed to detect in our study. The primary conclusions of our study were that (1) on the basis of segregation in a large Bedouin kindred with flanking microsatellite markers, hemolytic uremic syndrome can be inherited as an autosomal recessive disease tightly linked to the factor H locus, and (2) the affected Bedouin patients have abnormal cellular transport of factor H. These conclusions remain unchanged and are, in fact, strengthened by the excellent work of Buddles et al. We agree with their conclusion that further investigation of genotype-phenotype correlations in factor H abnormalities are needed to answer a number of interesting questions.

VAL C. SHEFFIELD,[1] LIHUA YING,[1] RIVKA CARMI,[2] AND DANIEL LANDAU[2]

[1]*Department of Pediatrics, University of Iowa, Iowa City; and* [2]*Soroka and Barzilai Medical Centers, Ben Gurion University of the Negev, Beer Sheva, Israel*

Address for correspondence and reprints: Dr. Val C. Sheffield, Department of Pediatrics, University of Iowa, 2615 JCP, Iowa City, IA 52242. E-mail: val-sheffield@uiowa.edu

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