Correspondence

reticular system. These findings could help in the
differentiation of the similar histiocytic disorder of
Letterer-Siwe disease. Chromosomal analysis should
be extended to include bone marrow cells and the
histiocytic cell line. The demonstration of an XX cell
line in a male patient would obviously be strong evidence
of chimerism. These studies might also help to clear up
the doubt about the malignant nature of the histo-
cytes.

It has been suggested (Buist et al., 1971) that the
disease may be a recessively inherited malignancy. The
evidence is from the occurrence of this rare disease in
sibs and first cousins (Goodall, Guthrie, and Buist,
1965). It may be of relevance that the patient in this
latest report was born of a cousin marriage.

J. S. WAINSCOAT

Department of Experimental Pathology,
The Medical School,
Birmingham B15 2TJ.

I. M. HANN

Alder Hey Children’s Hospital,
Liverpool L12 2AP.

REFERENCES

Blennow, G., Berg, B., Brandt, L., Mesetter, L., Löw, B., and
of chimerism? Archives of Disease in Childhood, 49, 960.

haemophagocytic reticulosis in first cousins. Archives of
Disease in Childhood, 46, 728.

Githens, J. H., Muschenheim, F., Fulginiti, V. A., Robinson, A.,
lymphoid chimerism secondary to probable maternalfetal

Goodall, H. B., Guthrie, W., and Buist, N. R. M. (1965). Familial
haemophagocytic reticulosis. Scottish Medical Journal, 10,
425.

Kadowaki, J., Thompson, R. I., Zuelzer, W. W., Wooley, P. V.,
chimerism in congenital immunological deficiency with
thymic alymphoplasia. Lancet, 2, 1152.

Dr. G. Blennow comments as follows:

Drs. Wainscoat and Hann point to three observations
that they consider are against a state of chimerism. We
would like to comment on their remarks.

(1) In the case of a graft rejection, the peripheral
blood lymphocytes are expected to be of host origin as
in our patient.

(2) One characteristic feature of haemophagocytic
reticulosis is remarkable increase in phagocytic reticulo-
endothelial cells in various organs. It is possible that
grafted, antibody-coated Rh-negative erythrocytes were
rapidly eliminated by these phagocytic cells and there-
fore escaped detection in the mixed field tests.

(3) The antiglobulin test was negative. As mentioned,
grafted cells may well be rapidly phagocytosed in vitro.
However, the results of the erythrophagocytosis ex-
periments in vitro may indicate that a few red cells
coated by antibody were actually present in the patient’s
peripheral blood.

We cannot therefore agree that the three observations
mentioned are definitely against a state of chimerism.
In our report it was not claimed that chimerism was
proven in our patient. However, we suggested that
future cases of haemophagocytic reticulosis should be
examined with such a possibility in mind. As pointed
out by Drs. Wainscoat and Hann, karyotype analyses
of the bone marrow would then be of great importance.

GÖSTA BLENNOW

Department of Paediatrics,
University Hospital,
S-221 85 Lund, Sweden.

Phenylalaninaemia

Sir,
The article on phenylalaninaemia by Blaskovics,
Schaefller, and Hack (1974) is timely in that it empha-
sizes the need for careful evaluation of all suspected
cases of phenylketonuria; some patients may be in-
danger if treatment is too restrictive, others may not
need any dietary treatment. The rather complex
classification proposed by the authors receives some
support from their own observations, but may need to be
modified when improved techniques permit more
accurate studies of enzyme activities. We prefer the
simpler and more practical classification given by Scriver
and Rosenberg (1973). They cover 5 types described
by Blaskovics et al., in three groups. (1) ‘Classical’
phenylketonuria. (2) Phenylketonuria (‘mild’ variant
with relaxed phenylalanine tolerance). (3) Phenylke-
tonuria (‘transient’ variant).

This grouping is more helpful in the clinical context.
The authors stress the importance of estimating the
daily phenylalanine tolerance of patients on treatment;
in the variant forms this is usually over 500 mg/day.

We estimated the daily phenylalanine intake required
by 24 of our patients on a low phenylalanine diet at
some point between the ages of 11 and 13 months, and
at a time when the serum phenylalanine was at an
acceptable level. In 18 cases the intake was between
275 and 450 mg/day (mean 365, 2 SD 120). They are
all, to the best of our knowledge, cases of classical
phenylketonuria. 5 patients were able to tolerate 500
to 600 mg phenylalanine/day. 4 of these returned to
normal diet between the ages of 1 year 3 months and
3 years. On normal diet one patient maintains a serum
phenylalanine level of 2–3 mg/100 ml, in the other 3
the range is 7–13 mg/100 ml. These 5 children are all
developing normally. All show impaired phenylalan-
ine tolerance on a loading test, giving curves above that
expected from a heterozygote, but below that of a
child with classical phenylketonuria. Perhaps none
needed treatment, yet at the time of diagnosis all but
one had a serum phenylalanine above 20 mg/100 ml
and metabolites of phenylalanine were detected in the
urine. The fifth ‘atypical’ patient tolerated exactly
500 mg phenylalanine/day but was maintained on a
diet for 7 years because she had 2 mentally retarded
sibs.

Our present practice is to start treatment if the serum