

which can be dismantled simply. In *Medicine and the State* these extracts are used to illustrate the author's theory that there is apparently a dark plot to say that the National Health Service is beyond all reproach. Such a contention is, of course, obvious nonsense to anyone who has any knowledge of affairs in this country, and needs no denial, but my experience seems to bear out your observation that "Dr. Lynch and Dr. Raphael . . . have culled from the world's medical literature references in support of a preconceived hypothesis".

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GORDON MCLACHLAN.

QUALIFICATION AS CONSULTANT

SIR,—“Unqualified Consultant” (Oct. 19) does well to introduce the plight of the S.H.M.O.s into this correspondence, since their existence is a continuous source of embarrassment in all discussion of the status of those engaged in senior hospital appointments in the N.H.S., and it will be well when the S.H.M.O. problem has been solved. It is unfortunate, therefore, that he oversimplifies the problem by defining a consultant as “a doctor holding a (consultant) post and not one with recognised qualifications”. Many S.H.M.O.s are filling posts which are designated as consultant posts in pursuance of para. 4 of R.H.B.(50)96. They are paid at S.H.M.O. rates (with the addition, since 1959, of a supplement in recognition of this).

In contrast, it was made clear, when the gradings were being reviewed in 1951, that anyone personally upgraded to consultant status at that time would be paid only at S.H.M.O. rates if holding a post classified as S.H.M.O.

The difficulty about the S.H.M.O.s did not arise from the absence of a criterion of recognised qualifications, as one might surmise from your correspondent's letter. It arose because in the Terms of Service of Hospital Medical Staff (June 7, 1949) it became possible for employing authorities to establish such appointments at less than consultant rates, and to keep them at these rates after a later survey had determined that the appointment was of a consultant nature.

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HYPOGLYCAEMIA IN THE NEWBORN

SIR,—Dr. Brown and Dr. Wallis (June 15) and Dr. Neligan and coworkers (June 15) conclude that intra-uterine malnutrition and “dysmaturity” predispose to this obscure condition. This is in keeping with a suggestion I put forward in 1956.¹

It has been shown that under normal conditions the enzyme glucose-6-phosphatase appears late in embryonic liver cells.² Up to this time foetal blood-sugar is supplied by the placenta, which contains this enzyme. Occasionally hypoglycaemia of newborns can probably be accounted for by retarded development of glucose-6-phosphatase in the liver. Persistence of this state beyond infancy represents glycogenosis. The survival of some infants with spontaneous disappearance of manifestations of the disease suggests that the retarded enzymatic development is speeded up in these cases. McQuarrie's cases of idiopathic spontaneously occurring hypoglycaemia in infants³ are probably examples of delayed transition from an embryonic to the extrauterine state of enzymatic activity.

This infantile hypoglycaemia is purely functional in character, varies greatly in severity from time to time, is unaccompanied by any demonstrable lesion (absence or near absence of α cells in the pancreatic islets was found in only 2 infants), and tends to

improve spontaneously. It is not found in older children. Convulsive seizures, however, indicate brain damage by hypoglycaemia.

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JULIUS BAUER.

HEAD CIRCUMFERENCE OF CHILDREN WITH PHENYLKETONURIA DURING DIETARY TREATMENT

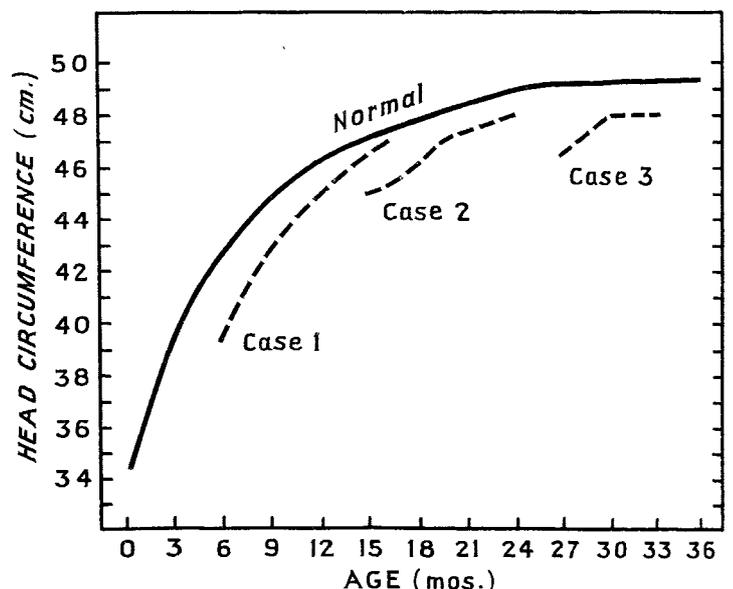
SIR,—The metabolic defect in phenylketonuria is known to be caused by absence of active phenylalanine hydroxylase. How the metabolic defect causes mental retardation is as yet unknown, but when phenylalanine intake is restricted early the child may develop normally.^{1,2} Better results are obtained when treatment is begun before the age of six months than at the age of two years.³ After the initiation of well-controlled dietary therapy we usually find that the biochemical abnormalities disappear, the child's behaviour and electroencephalogram improve, the hair darkens, and eczema may disappear.

There is another result of dietary therapy that has not been reported so far. In 3 patients we saw a rapid growth of the skull during treatment with a phenylalanine restricted diet. The serum phenylalanine levels stayed in the normal range most of the time. They were determined by the method of Van de Heuvel-Wadman,⁴ the blood-samples being drawn early in the morning after eight to ten hours of fasting. The circumference of the head was measured at intervals of three months and always by the same person. Each value is the average of three consecutive measurements.

A boy (case 1) was diagnosed as having phenylketonuria and a phenylalanine restricted diet was begun at the age of six months. At that time the head circumference was 40.3 cm. (normal at this age according to Harvie⁵ 43.8 cm.). After less than a year on the diet the skull had grown to practically normal size (see figure). Meanwhile his development quotient had gone up from 40 to 70 (Bühler-Hetzer test).

Another boy (case 2) was treated for phenylketonuria from the age of fifteen months. During therapy his mental development quotient did not improve. His head circumference, however, increased from 46 to 49 cm. in nine months, whereas the normal increase at that age is from 48 to 49 cm.

1. Horner, F. A., Streamer, C. W. *Amer. J. Dis. Child.* 1959, **97**, 345.
2. Centerwall, W. R., Centerwall, S. A., Armon, V., Mann, L. B. *J. Pediat.* 1961, **59**, 102.
3. Berman, P. W., Graham, F. K., Eichman, P. L., Waisman, M. A. *Pediatrics*, 1961, **28**, 924.
4. Van der Heuvel, J. M., Wadman, S. K. *Clin. Chim. Acta*, 1960, **5**, 824.
5. Harvie, F. H. *Pediatric Methods and Standards*. Philadelphia, 1962.



Head circumference in phenylketonuria after phenylalanine restriction.

1. Bauer, J. *The Person behind the Disease*. New York and London, 1956.
2. Villee, C. A. *J. appl. Physiol.* 1953, **5**, 437.
3. McQuarrie, J. *Amer. J. Dis. Child.* 1954, **87**, 399.

In a third boy (case 3) phenylketonuria was diagnosed at the age of two years and three months and treatment was started at the same time. His mental development did not progress either. During the first three months of therapy his head circumference increased from 47.3 to 49 cm. Normally it would not increase at all during this period.

As could be expected, treatment produced the best results in the six-month-old boy.

We conclude that microcephaly is never a contra-indication to a therapeutic trial in this disease, certainly not at an early age.

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THALIDOMIDE AND THE UMBILICAL ARTERY

SIR,—We have examined the umbilical cord of 4 cases of thalidomide embryopathy in which the infant died shortly after birth.

A single umbilical artery was found in 2 of these 4 cases, an attenuated right umbilical artery was present in 1, and in the remaining case the umbilical arteries were normal. In each case with a single umbilical artery, the iliac and femoral arteries of the side where the umbilical artery was missing were about one third of their normal size, and the intra-abdominal umbilical artery of the same side was lacking. In case 3, in which the right umbilical artery was half its normal size, the intra-abdominal umbilical artery of the same side was also attenuated, though the right iliac and femoral arteries were normal.

These 3 cases with abnormalities of the umbilical artery had multiple congenital malformations of the internal organs, including the heart and genitourinary system, besides limb deformities. By contrast, the case with normal umbilical arteries showed no malformations of the internal organs.

Details of the 4 cases are given in the accompanying table.

Studies of the single umbilical artery^{1,2} show increased association of a range of congenital malformations with this circumstance. The 17-18 syndrome has been reported to accompany single umbilical artery.³⁻⁶ But an extensive search of published reports on thalidomide embryopathy has revealed no case with a single umbilical artery except one described by Dunn et al.⁷ in which there was binovular twin pregnancy. This case presented phocomelia of the upper limbs and a pilonidal sinus. A single umbilical artery is present in 7% of twin pregnancies, compared

with an overall prevalence of 1%, and it is hard to tell whether in this case the abnormality is attributable to twin pregnancy or to thalidomide. But our finding of abnormalities of the umbilical arteries in 3 out of 4 cases of thalidomide embryopathy suggests that the umbilical cord of every thalidomide baby should be examined for such abnormalities.

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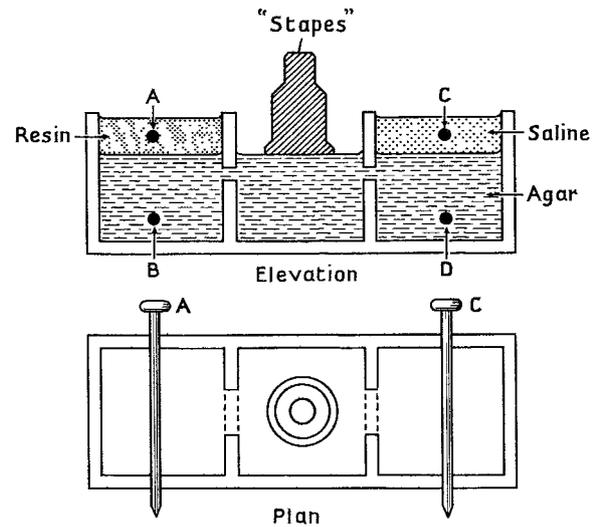
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CHARACTER OF THE TECTORIAL MEMBRANE

SIR,—The results of our analysis of the tectorial membrane¹ have prompted us to make a model imitating part of the energy-transducing mechanism of the cochlea.

The model consisted of an agar-gel with electrolyte overlaid by a liquid ion-exchange resin not miscible with water.



Polyethylene cells with agar-gel and overlaid resin or saline solution; A, B, C, and D are indifferent electrodes made of stainless steel rigidly fixed in walls.

(‘Amberlite’, LA-1, British Drug Houses.) The resin was diluted with organic solvent, miscible with both water and resin phases, and saturated with water containing the same electrolytes as in the agar-gel. This system was left for a little time to allow the equilibrium distribution of electrolytes for the system to become established. Indifferent electrodes were inserted rigidly into both phases (see accompanying figure).

The resin-gel system was contained in a small polyethylene

1. Naftalin, L., Harrison, M. S., Stephens, A. *Lancet*, 1963, i, 1192.

DATA ON 4 CASES OF THALIDOMIDE EMBRYOPATHY

Case no.	Date of birth	Sex	Gesta-tional age (weeks)	Weight at birth (g.)	Age at death (days)	Complica-tion of pregnancy	Limb malformations	Associated malformations	Thalidomide	Umbilical arteries
1	Sept. 8, 1962	M	35	2100	Still-born	Hydram-nios	Absence of both radii, ulnæ, and right humerus; left humerus short stumps of bone; 3 fingers on each side	Interventricular septal defect; bilateral abdominal testicles; very mobile cæcum and ascending colon	Taken	One (left)
2	April 2, 1963	M	37	2200	16	None	Amelia of arms; phoco-melia of legs; 6 toes on right, 7 on left	Bilateral anophthalmos; anomalous lobulation of right lung; left hydronephrosis; very mobile cæcum and ascending colon	Sedative taken, but nature unknown	One (right)
3	June 9, 1962	F	42	2800	5	None	Phocomelia of arms and legs; 3 digits and 6 toes on each side	Hæmangioma of upper lip; depressed nasal bridge; interauricular septal defect; abnormal lobulation of lungs; absence of appendix	Taken	Two (right attenuated)
4	May 4, 1962	F	43	2300	14	None	Single arm bone on each side; 3 digits on right, 4 on left	Slight deformity of left ear; pilonidal sinus	3 sorts of seda-tives taken but nature unknown	Two