A Method for Feeding the Phenylketonuric Infant

BARBARA CLAYTON,* PH.D.ED., M.D.; DOROTHY FRANCIS,* S.R.D.;
Sir ALAN MONCRIEFF,* C.B.E., M.D., F.R.C.P.

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Though the dietary treatment of phenylketonuria has been described in a Report to the Medical Research Council of the Conference on Phenylketonuria (*Brit. med. J.*, 1963, 1, 1691), numerous requests for advice have been received at the Hospital for Sick Children, Great Ormond Street. We consider that a simple statement of how we actually feed such an infant at this hospital may be of help to other people.

Phenylalanine occurs in proteins which provide the nitrogen for the body. In the treatment of phenylketonuria most of the nitrogen is provided by synthetic foods containing negligible amounts of phenylalanine: Minafen (Truefood Ltd.) for infants, and Cymogran (Allen & Hanbury Ltd.) for toddlers and children. Since phenylalanine is essential for growth some must always be provided by giving milk to infants, and, later, other foodstuffs in carefully calculated amounts. If insufficient is given the infant will fail to grow, develop rashes, and may die.

Low-phenylalanine Diet for an Infant

1. Weigh the infant.
2. Calculate the amount of Minafen: 1 oz. Minafen/kg. body-weight/day; or $\frac{1}{4}$ oz. Minafen/lb. body-weight/day.
3. Calculate the amount of milk required daily: 1 oz. milk/kg. body-weight/day (providing approximately 50 mg. phenylalanine/kg. body-weight/day); or $\frac{1}{4}$ oz. milk/lb. body-weight/day (providing approximately 25 mg. phenylalanine/lb. body-weight/day). Phenylalanine is an *essential* amino-acid, and milk must *always* be given with Minafen, even when the diet is first begun. The amount of milk will need adjusting for each infant. (See 6.)
4. Calculate the amount of fluid required daily: 5 oz. fluid/kg. body-weight/day; or $2\frac{1}{2}$ oz. fluid/lb. body-weight/day. The feeds are then given four-hourly, and each feed *must* contain a portion of the day's milk.
5. Give the daily vitamins: 5 ml. Ketovite Supplement Syrup (Paines & Byrne Ltd.) daily; and vitamin C—e.g., orange juice, rose-hip syrup.
6. Phenylalanine must be estimated in the blood twice weekly until the blood level remains normal, and thereafter at weekly intervals. (It is *essential* to have a rapid laboratory service. Urine testing must *never* be used for controlling the diet.) Depending on the phenylalanine level, adjust the amount of milk as follows: If the blood level is 5 mg./100 ml. or higher, remove half to 1 oz. milk. If the blood level is 4, 3, or 2 mg./100 ml.—no change in milk ration. If the blood level is 1 mg. or less/100 ml.—add half to 1 oz. milk.
7. Increase the allowance of Minafen as the infant gains weight.

Weaning the Phenylketonuric Child

1. Wean at the same stage as in a normal infant.
2. Always check the blood phenylalanine level and see it is satisfactory before introducing a new food.
3. Teach the infant to use a spoon or cup by giving orange juice, rose-hip syrup, or blackcurrant juice from these. Introduce puréed fruit (not banana), gradually increasing to 2–3 tablespoonfuls.
4. Remove part of the milk from the 10 a.m. feed, and use it to make a mash with "Liga Low Protein Rusks."¹ Gradually

use all the 10 a.m. milk for this mash. Additional rusks may be mashed with fruit juice or water if desired. If the mash is not all eaten the milk in it must be replaced and given to the infant as such.

5. Remove the milk allowance from the 6 p.m. feed and make it into a pudding with cornflour, sugar to sweeten, and some water if desired. If the pudding is not all eaten the milk must be replaced.

6. Introduce purée vegetables before the 2 p.m. feed, and to this may be added Kosher or pure vegetable margarine or tomato juice. (*N.B.* Do *not* give peas, beans, spinach or potato.)

7. Gradually remove the milk from the 2 p.m. feed and instead give boiled potato: 2 oz. potato=1 oz. milk. Start with a little potato—i.e., $\frac{1}{2}$ oz., which is equal to $\frac{1}{4}$ oz. milk—and give the rest of the milk separately.

8. As soon as the infant can chew (a) offer low protein Liga rusks or rusks made from wheat starch, pure vegetable margarine, and sugar; (b) offer raw apple or pear; (c) give chopped foods instead of purées.

9. The 10 p.m. and 6 a.m. feeds can be omitted when the infant will take all the Minafen ration at the other three feeding times. By this stage, the quantity of Minafen will be considerable, and a change to Cymogran is indicated. Phenylalanine levels *must* be estimated twice weekly during the change-over period.

Change from Minafen to Cymogran

1. Calculate the amount of Cymogran: $\frac{1}{4}$ oz. Cymogran/kg. body-weight/day; or $\frac{1}{4}$ oz. Cymogran/lb. body-weight/day. Give it as three feeds before each meal.
2. Calculate the amount of phenylalanine on which the child has already been stabilized, taking into account the phenylalanine contributed by the protein substitute: Minafen contains 6 mg. phenylalanine/oz. Cymogran contains 3.3 mg. phenylalanine/oz. Give the phenylalanine allowance equally between the three meals.
3. Give the vitamins required with Cymogran: 3 tablets Ketovite (Paines & Byrne Ltd.) daily, and 5 ml. Ketovite Supplement Syrup daily.
4. Give additional calories from foods of low phenylalanine value—e.g., fruit, vegetables, low-phenylalanine bread.
5. Increase the Cymogran as the child gains weight.
6. Once the child is stabilized on the Cymogran regimen weekly and later monthly phenylalanine levels should be estimated.

Notes

- (a) The above regimen is satisfactory up to the age of 1 year.
- (b) With infections serum-phenylalanine levels rise, the child may not eat, and careful stabilization is again necessary.
- (c) The phenylalanine content of many foods is given in the *British Medical Journal* (1963, 1, 1691).
- (d) The following tables are available on request from the Dietitian, the Hospital for Sick Children, Great Ormond Street, London W.C.1: (1) Foods which are forbidden. (2) Foods which are allowed freely. (3) Exchanges for milk: portions of foods containing 50 mg. phenylalanine. (4) Portions of fruits and vegetables containing 20 mg. phenylalanine. Suitable recipes for bread, biscuits, puddings, etc., are also available.
- (e) The following may be prescribed on E.C. 10: (1) Minafen and Cymogran. (2) The vitamin preparations. (3) Energen wheat starch for cooking and making bread.

* The Hospital for Sick Children, Great Ormond Street, London W.C.1.
¹ Liga Food Co., 23 Saxby Street, Leicester, England. One rusk contains 1.4 mg. phenylalanine and supplies 50 calories.