No real difficulty was encountered in the administration of terramycin. Toxic effects were not observed.

We are indebted to Dr. Ivor Marsh, late casualty officer, Belgrave Hospital for Children, for his interest and enthusiasm; to the Medical Research Council Antibiotic Clinical Trials (Non-tuberculous Conditions) Committee; and to Dr. R. K. Phillips, of Pfizer, Ltd., for supplies of terramycin.

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Medical Memoranda

Familial Cystinuria

Cystinuria is an inborn error of metabolism in which the amino-acid cystine, together with lysine and arginine, is excreted in greatly increased quantity in the urine. There is also evidence that some cystine is excreted as a labile precursor of unknown composition which liberates cystine on standing. (In the absence of calculi, cystinuria is a symptomless condition.) The output of cystine is not increased by feeding this amino-acid, as it is excreted as a sulphate. However, if cysteine or methionine is fed there is an increased output of cystine. High-protein diets would therefore seem inadvisable. The factors responsible for keeping cystine in solution in the urine are not yet known, but calculus formation does not seem to be directly related to the concentration of cystine in the urine. It has been reported that solution of stones will take place if the urine is kept at a pH of 7.5 by administration of sodium citrate.

The occurrence of cystinuria in families has recently been reviewed by Dent and Harris (1951). In addition to the 11 families reported in the literature they have collected a further 11. As a result of their study they have put forward the view that cystinuria occurs in three types of case: (1) in the Fanconi syndrome, associated with many other amino-acids; (2) in Wilson's disease (hepato-lenticular degeneration); and (3) "classical" cystinuria, the type considered here, in which there is also an increased excretion of lysine and arginine: several of the cases of this type in the literature were, in their opinion, examples of the Fanconi syndrome.

In the past the detection of cystinuria depended mainly on the recognition of the typical hexagonal crystals of cystine in the urine or on the qualitative cyanide nitroprusside test. Lewis (1932) examined the urine of 10,534 students by the cyanide-nitroprusside test and found an increased quantity of cystine to be present in 1 in 600. It seems likely that the use of the newer and more sensitive methods of chromatography and polarography will bring more cases to light.

FAMILY HISTORY

The family reported here was discovered initially through the finding of cystine crystals in the urine of one member. Subsequent examination of the urine of all available members of the family by the cyanide-nitroprusside test and the methods of paper chromatography and polarography brought one further case to light.

○ URINE CYSTINE ABSENT ● CYSTINE PRESENT ◐ NOT EXAMINED

The family tree, showing ages of members and presence or absence of cystine in the urine.

Case 1.—A retired coach-driver aged 51 was operated on for removal of bladder stones at the ages of 6, 14, and 25 years. He had a right nephrectomy for "staghorn" calculus in 1943. He was admitted to St. Mary's Hospital in January, 1951, for treatment of cardiac failure following myocardial infarction in May, 1948. On routine treatment he complained of a burning pain on micturition. Microscopical examination of the urine revealed cystine crystals. The cyanide-nitroprusside test was positive and paper chromatography showed an increased excretion of cystine, lysine, and arginine. An x-ray picture of the bladder showed two stones.

Case 2.—A younger brother (aged 42) of Case 1. There is no history of renal or cystic calculus. His only symptoms are those due to a peptic ulcer. Paper chromatography showed the same abnormal pattern of amino-acid excretion as in Case 1.

In both these cases polarographic studies (Reed, 1942) suggested that the output of cystine was about 0.5 g. a day. The range of output in normal persons is 40–80 mg. in 24 hours. In cystinurics the range has been given as 0.4–1.8 g. in 24 hours.

The investigations of Dent and Harris tend to show that classical cystinuria is inherited through a simple recessive factor or is a heterozygous condition with irregular manifestation. The family history reported here would fall into either of these forms of inheritance.

I am indebted to Dr. J. W. Litchfield for permission to publish this case report, and to Dr. H. Harris and Professor F. L. Warren for examining the urine by chromatography and polarography at University College, London.

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Blackwater Fever Treated with Cortisone

CASE REPORT

A Yoruba boy aged 19 months suddenly became ill at 11 a.m. on August 25, with a high fever, and was given an injection of quinine before admission to hospital the same day. He had been quite well, apart from having attacks of fever at varying intervals since birth, for which he had always received quinine. This treatment had apparently terminated the fever rapidly on each occasion.

On admission he was in a collapsed condition; his temperature was 104° F. (40° C.), his pulse 140 and of a poor volume, but he appeared to be well nourished. The mucous membranes were very pale and no icterus was noticed at the time. The heart was apparently normal. In the lungs scattered sibili were audible. The spleen was palpable three fingerbreadths below the costal margin, and the edge of the liver was just palpable. At 11.30 a.m. he passed 10 ml. of almost jet-black viscid urine, and methaemoglobin was demonstrated spectroscopically. The haemoglobin was 4.8 g.%, and a blood film showed large numbers of *Plasmodium falciparum* trophozoites. On August 26 he appeared to be moribund. The conjunctivae were icteric, and his general condition had deteriorated. By 3.45 p.m. only 1 oz. (28 ml.) of black urine had been passed since the last specimen was collected at 11.30 a.m. on the previous day.

Cortisone, 100 mg., was given intramuscularly, and at 8 a.m. on August 27 there was a noticeable clinical improvement: the child sat up and ate; his temperature was 97.2° F. (36.2° C.), and his pulse 94. He had taken 70 oz. (2 litres) of fluid in the past 24 hours and was known to have excreted 8½ oz. (240 ml.) although he had been incontinent since 3.45 p.m. on August 26. The first three specimens following the injection of cortisone were dark, but were lighter than previous specimens, and the next three

specimens appeared normal in colour. Cortisone, 100 mg., was again given intramuscularly. This was followed by a dose of 50 mg. on August 28 and 29. On the 30th he was sitting up; the conjunctivae were icteric, and there was slight oedema of the feet and face. No further cortisone was given.

The urinary output on August 28, 29, and 30 was about 20 oz. (570 ml.) a day, and some urine was voided into the bed; it contained no methaemoglobin. The anaemia was treated with iron, and the malaria from the outset with mepacrine. The child made an uneventful recovery and was discharged.

COMMENT

It is suspected that blackwater fever is a haemolytic disease occurring in those infected with *P. falciparum* and whose cells have been sensitized by quinine. As cortisone lowers the sensitivity of body tissues in certain allergic states (Carey *et al.*, 1950), it was given in the hope that it might reduce the sensitivity of the cells to the circulating haemolysin and relieve the renal cortical ischaemia responsible for the anuria.

Although the clinical state of cases of blackwater fever is known to alter rapidly, it was felt that the condition of this child on the second day was such that a fatal outcome was inevitable. Cortisone seemed to have had an effect in this case.

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Advantages of a Perineal Curve Applied to Midwifery Forceps

The first patterns of midwifery forceps—those of Chamberlen in the seventeenth century and of Edmund Chapman (1733) and William Giffard (1734)—were short and straight. William Smellie (1752) recommended the short straight forceps for general use, and he it was who discovered, to his great delight, how this instrument could be used for rotating the head in occipito-posterior presentations. Nevertheless, it was Smellie, too, who lengthened the forceps and added the pelvic curve in order to reach the head higher up in the pelvis—an invention also claimed for André Levret (1751) and Benjamin Pugh (1754). The pelvic curve was also an outstanding feature of the *mains de fer* designed by Palfyn in 1720. In the course of time the long curved forceps has supplanted the straight forceps for general use.

It was soon discovered that there were several disadvantages in the pelvic curve, notably the fact that traction on the handles is inefficient, as the direction of force is not in the axis of the birth canal. Many attempts to overcome this difficulty were made, and it was finally solved by Tarnier in 1877 with the axis-traction forceps. By the time axis traction became definitely established the necessity for the high forceps operation had vanished with the introduction of the relatively safe forms of caesarean operation in use to-day. The principle of axis traction still holds, but the patterns of forceps in general use are unnecessarily heavy and powerful for the low forceps operation, and many obstetricians have abandoned their use. Furthermore, with the long forceps with the pelvic curve, whether with axis-traction rods or without, it is not possible to obtain a good cephalic application when the head, in mid-pelvis or higher, lies in the transverse position. Severe lacerations of the birth canal have followed attempts to use the long curved forceps to rotate the head in this position, and most obstetricians in this country condemn the practice.

As long ago as 1769 R. Wallace Johnson, realizing some of the disadvantages of the long curved forceps, endeavoured

to overcome them by bending back the shanks in a perineal curve, thus bringing the handles more into line with the axis of the blades. This was the first of many attempts to obtain axis traction without the use of additional apparatus. The well-known forceps designed by Kielland (1916) also has a perineal curve for the same purpose, and solves most of the problems of applying forceps to cases of deep transverse arrest, but it has not yet become very popular, probably because of certain other features which require a special technique to be practised if this instrument is to be used as Kielland intended.

During the winter of 1949–50 I had an old forceps of the Ramsbottom type replated, and took the opportunity of getting the shanks bent back at an angle of about 30 degrees, close to the lock (see Diagram). I have used this instrument now for nearly three years, and have found that the alteration exceeded my expectations in making the forceps more efficient and handy. A line drawn through the ends of the handles and passing through the centre of the fenestrae almost exactly corresponds in direction with a line drawn in the same way but passing through the traction crossbar of the Barnes–Neville axis-traction forceps I used formerly. The curve of the instrument as seen from the side very closely corresponds with the curve of the Kielland forceps.

I claim that the addition of a perineal curve in this way gives the following advantages over the ordinary long forceps: (1) It provides axis traction for all low forceps deliveries without the addition of any traction rods, and nearly as efficiently as that provided by the Barnes–Neville forceps. (2) It can be more easily applied to the foetal head in transverse positions, giving a proper cephalic grip. It could be used for rotating the head in much the same way as the Kielland forceps is used, though I have not yet tried it. (3) It is more convenient than the normal type for use on the aftercoming head in breech presentations, as the handles are bent away from the body of the child in the same way as those of Piper's forceps (Piper and Bachman, 1929). (4) It is an ideal instrument for the general practitioner, as it is more versatile and more convenient to carry.

I claim no originality in this instrument, the idea being nearly two centuries old. Peterson (1952) has described a very similar forceps, which he has used for over 100 deliveries with complete satisfaction, and claims much the same advantages for it. It is my view that a special instrument is not necessary, and that it will suffice to give a perineal curve to any standard instrument, such as Simpson's or Anderson's.

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