A CASE OF ENTEROGENOUS METHAEMOGLOBINAEMIA (?CONGENITAL) IN A CHILD

BY

REGINALD MILLER, M.D., F.R.C.P.

Physician, Paddington Green Children's Hospital, London.

Enterogenous methæmoglobinæmia, a comparatively rare disorder in adults, has never, so far as I am aware, been described in a child, although Prof. L. G. Parsons¹ has recorded a case of sulphæmoglobinæmia in an infant of nine months old. For this reason the following case is recorded.

P.B., female, aged 9 years. She was first seen in April, 1927, when she was brought to the out-patient department at the Paddington Green Children's Hospital for bronchitis. Her obvious blue colour was explained as being due to congenital heart disease, but as it did not resemble the cyanosis of that condition, she was admitted for investigation.

The girl had lived in an institution for the past two years and it was impossible to get into touch with her mother or father. From the age of nine months until she was admitted to the institution, she was under the charge of a foster-mother. This woman when interviewed stated that the child was blue when she first took charge of her, and that she had always understood that the blue colour dated from birth and was due to congenital heart disease. The colour had remained the same all the years she had had charge of the child, and it had not altered while in the institution from which she was brought to hospital.

The girl's health was stated to have been good except for obstinate constipation and a tendency towards abdominal distension. Although it was understood that her colouration was due to cardiac abnormality, it was realized that apart from this one symptom the heart's condition seemed to do no harm to the child.

FIRST ADMISSION. Admitted in April, 1927, the girl was well grown and developed for her age (9 years). Her face and body were plump. Her hair was light brown, and her eyes pale blue. Apart from her cyanosis she appeared in good health. She was, however, persistently and severely constipated, and her abdomen was rather distended but only to an extent suggestive of constipation.

The blue colour of the child was very obvious at any time and was increased when she became flushed or hot. The tint of the blueness was not the same as that of cardiac cyanosis, having a more leaden or steel tinge in it. The extremities, although blue, were quite warm; the tips of the ears were not cyanosed. The white skin, as of the abdomen, was of a colder white than in an ordinary child.

The heart showed distinct enlargement of the left ventricle. The apex-beat was $\frac{3}{4}$ -inch outside the nipple line and was rather thrusting in character. A skiagram confirmed this. The pulse was good. There was no trace of dyspnœa on exertion. There was no clubbing of the fingers. The blood showed no erythrocytosis. The red cells numbered on several counts between $4\frac{1}{2}$ and $4\frac{3}{4}$ millions. The leucocytes numbered about 6,500; and the hæmoglobin which was estimated with difficulty owing to the peculiar tinge of the blood, stood at about 80 per cent. The colour index was .9.

The abdomen was full and rather distended, but to no very remarkable degree. Nothing abnormal could be detected on palpation. An opaque enema showed that the colon was large and easily distended, but was nothing like the voluminous colon of megacolon. An enema of two pints was very easily delivered and retained by the child. Without treatment, the bowels never acted. The stools showed no signs of undigested food or colitis, but were rather abnormally offensive. Bacteriologically there was at first found a great excess of streptococci of the common faecal type over *B. coli*, but after a course of colon lavage this excess largely disappeared. The 'nitroso-bacillus' could not be found in three examinations. The only abnormality in the urine was the very large amount of indican which was constantly present. The child's temperature was regularly raised to 99.5° or 100° at night.

A spectroscopic examination of the blood was kindly carried out for me by Dr. W. H. Hurlley at St. Bartholomew's Hospital who diagnosed the condition as one of methæmoglobinæmia on the following grounds:—

(1) A diluted solution of the blood showed a band to the left of the D line, at '3' on an arbitrary scale.

(2) This band disappeared when the blood was reduced with a trace of sodium hydrosulphite. (This is typical of methæmoglobin: the band of sulphæmoglobin would remain.)

(3) On the addition of ammonium sulphide the band at '3' instantly disappeared, while a band at '3.5' appeared slowly. (This is the band of sulphæmoglobin. The '3.5' mark is nearer the D line than is the '3' mark.)

(4) Sulphuretted hydrogen was passed into the diluted blood. This caused the '3' band to disappear immediately, and a sulphæmoglobin band at '3.5' to appear rapidly in its stead.

It was assumed that the three signs, methæmoglobinæmia, indicanuria and persistent low fever were interdependent, and attempts were made by various measures to influence them during the three months the child remained in hospital. Chief reliance was placed upon the regular use of large enemata of double normal saline in alkaline solution. These were given daily over a long period. Various so-called intestinal disinfectants were also tried, and a three week's course of *B. acidophilus* milk was given, the cultures for which were kindly supplied by Dr Laurence Garrod. All these measures were quite unsuccessful in influencing any of the three symptoms already mentioned. The alterations in the child's colour were not more than were ultimately learned to be due to the flushing or the pallor of her skin. The indicanuria persisted in spite of the prevention of any apparent constipation by the use of enemata, and the low fever was also uninfluenced.

The girl was discharged after a stay of three months in hospital.

SECOND ADMISSION. The child was not seen again for nearly two years when she was sent for and re-admitted on March 8th, 1929. In the interval she had kept in her usual state of health and on re-examination no change in her condition could be found. Dr. Hurlley kindly re-examined the blood and confirmed his previous diagnosis.

At Dr. Morley Fletcher's suggestion daily colon douches of potassium permanganate (1 in 4,000) were started on March 23rd., and a careful watch was kept on the amount of indican in the urine. This began to lessen about April 8th, and an improvement in the child's colour was noted about ten days later. On April 30th, the urine became free of indican for the first time during all the weeks she had been under observation. It reappeared from time to time, but in small quantities only. The child's colour made a very definite improvement, so that when she was not flushed the abnormality of the complexion was hardly recognizable, but when heated by exercise her methæmoglobinæmic tint became again obvious. Improvement beyond this point could not be obtained. A course of completely vegetarian diet appeared to make no difference to the girl's progress.

As the child's colour improved it was noticed that her hair became lighter, giving rather the appearance of a child's hair which had been bleached in the sun. The variations in the indicanuria were observed to precede the variations in the colour of the child's complexion by about 24 to 48 hours. An increase in the indicanuria would be followed by an increase in the degree of cyanosis, and vice versa.

The girl was discharged on August 14th, 1929.

Summary.

(1.) A case of enterogenous cyanosis due to methaemoglobinæmia has been described in a girl first seen at the age of 9 years. The cyanosis was certainly present at the age of 9 months, and is said to have been present at birth. It was attributed to congenital heart disease.

(2.) Apart from the abnormal colour the girl appeared in good health. The absence of any symptoms of illness is perhaps to be attributed to the comparative absence of anæmia.

(3.) The heart showed an enlarged left ventricle : there was no erythrocytosis.

(4.) The methaemoglobinæmia was accompanied by obstinate constipation, excessive indicanuria and some persistent low fever at night. The degree of cyanosis varied directly with that of the indicanuria.

(5.) Considerable improvement, but by no means a complete cure, resulted from the daily use of enemata of potassium permanganate solution (1 in 4,000). Other measures appeared useless.

REFERENCE.

1. Parsons, L. G., *Goulstonian Lectures*, Lond., 1924, 50.