

SHEILA JONES (Mother--Mary Jones)
Born: October 1, 1949

Miles made on our film
phenylketonuric child

Parents separated. Roman Catholic. Third child in the family, the other two being normal. Address -- 33 / 3 Matlock Road, Tyseley, Birmingham.

Referred by Dr. Allin. ^{Demo.} 38 Mirfield Rd. ^{Solihull}

March 13/51 Sheila was first seen in the Out-Patient Department on this day, she was then 17 months old, could sit only, and rocked continuously. Her mother was asked to return with a specimen of urine in a week's time. She did not bring the specimen of urine. A letter was therefore sent to her with a container, and she returned this with urine to the Children's Hospital, and it arrived on April 14, 1951. Dr. Bickel tested the urine and it was positive for phenylpyruvic acid.

Apr. 28/51 Letter sent to Dr. Allin saying that "Sheila suffers from phenylpyruvic oligophrenia". "As far as we know no line of treatment is of any avail, but we will admit her to hospital in a month or two for further investigation".

Sept. 25/51 - Oct. 11/51 Sheila was admitted to hospital for observation. Urine from siblings was tested and was negative.

Nov. 21/51 - May 17/52 Sheila was admitted to hospital for the second time and on November 27th was put on a phenylalanine-free diet. On December 18th this was changed to a phenylalanine-poor diet. During the phenylalanine-free period, the phenylalanine plasma level fell from 5 mgm. per 100 ml. to normal (under 1.5 per 100 ml.). The phenylalanine excretion in the urine dropped from 500 mgms. per day to normal levels of under 10 mgms. per day. The phenylpyruvic acid excretion dropped from 500 mgms. per day to 38 mgms. per day. During this period, the child lost weight slowly but steadily, as was to be expected when an essential amino acid was omitted from the diet. The child was therefore changed to a phenylalanine-poor diet during which period the phenylalanine content of blood and urine and the phenylpyruvic acid content of the urine rose again, but was still only about one third of the original values. Under this regime, the patient maintained her weight, increasing it slightly towards the end of the period. She was sent home on a diet described in the diet sheet appended. This should provide her with no more than 1 gram

phenylalanine daily. It is still too early for an assessment of any mental improvement under the phenylalanine-poor diet. The mother is convinced that the child is lively, plays more, and altogether behaves more sensibly. The child is under weekly out-patient supervision.

After discharge from hospital she was to attend with her mother every week for the prepared formula.

Oct. 9/52 Mrs. Jones was given 7 bottles of treated casydrol, but on this occasion each bottle contained 5 grams of phenylalanine. Sheila was given her first dose on a Friday and by nightfall she was crying and banging her head just as she used to. On the following day she cried continually and her eyes looked heavy. She could no longer stand holding onto a chair and her crawling was weak. Her mother felt there must have been a mistake in the preparation of the diet and discontinued it. On the following Wednesday, October 15th, Mrs. Jones came for the first time without Sheila and in tears, because she said something had put Sheila back to where she had been when we first saw her, and that she was unable to stand and was quite lifeless. Two days later we visited the house, Sheila was now back on her proper diet and the two lodgers confirmed Mrs. Jones' story.

Oct. 23/52 We saw Sheila again. She is now well established on the proper regime and seemed back to normal.

Nov. 5/52 - Dec. 24/52 Sheila was admitted to the Children's Hospital for the third time, she was still obviously mentally defective, but she walked with support and laughed when tickled. On November 26th 4 grams of phenylalanine was added to the diet daily, and the following day 5 grams was added to the diet. She had a very disturbed, noisy night unlike any in hospital on this occasion and had to be sedated with chloral. On the following day she was extremely irritable and drowsy and vomited. Her condition remained essentially unchanged until the 3rd of December when she was once more put on the phenylalanine-low diet.

The last note we have of her was a letter from Dr. Blainey to Dr. Allin and this was written on July 28th, 1955.