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BOOK REVIEW

Sheila – Unlocking the Treatment for PKU

by Anne Green

Paperback, 192 pages
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This is the story of the early days of the recognition and efforts to treat the inborn error of metabolism, phenylketonuria (PKU). It is not a textbook of metabolic disorders. Rather, the focus is the life and family of Sheila Jones, the first child in whom dietary treatment was attempted. It is set in the context of the development of Birmingham Children's Hospital (BCH) and the scientific and medical people involved. The condition of mental impairment and excess phenylalanine in the blood and its products in the urine was recognised in the 1930s but in 1951, when Sheila was first referred, there was no known treatment.

Following the idea that a reduction in intake of phenylalanine might help, the BCH biochemists worked to create a suitable protein source. This led to recognition that treatment starting after the first few weeks of life was useless and thus, eventually, to the neonatal screening programme.

Following Sheila's progress provides an account of the changes and challenges in care for those with learning disabilities and the problem of transfer of care from paediatric to adult services.

Among collateral stories of medical and biochemical staff is that of Marion Hickmans, a woman who first graduated in Chemistry in 1905, set up a pioneering biochemical laboratory and still masterminded the process to develop the special diet in the early 1950s – a career, well worth the telling. The author is herself a biochemist who worked later in the same laboratory service.

Included, are many relevant illustrations and some useful diagrams and charts related to the condition. This account of the early days of recognition of PKU will appeal not only to sufferers and their families, medical historians and health professionals, but also to the legions of mothers whose have wondered about the neonatal screening of their newborn infants.

Sheila L B Duncan
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