Review Of "The PKU Paradox: A Short History Of A Genetic Disease" By D. B. Paul And J. P. Brosco

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In 1995, Diane Paul wrote an important book, most pediatricians will never encounter a case during the title? It is because even though PKU is rare and excellent. The prognosis for normal neurological functions is difficult dietary regimen that restricts the intake of the amino acid phenylalanine, an essential amino acid, the development of Mayo and PKU, led to strategies for trying to control the disorder by dietary intervention.

The PKU Paradox: A Short History of a Genetic Disease. Johns Hopkins Biographies of Disease. By Diane B. Paul and Jeffrey P. Brosco. Baltimore (Maryland): Johns Hopkins University Press. $24.95 (paper). xxv + 289 p.; ill.; index. ISBN: 978-1-4214-1131-6 (pb); 978-1-4214-1132-3 (eb). 2013. In 1995, Diane Paul wrote an important book, Controlling Human Heredity: 1865 to the Present (Atlantic Highlands (NJ): Humanities Press), about the eugenics movement. She explored the ethical, legal, and social issues posed by the rapidly emerging powers of medical and genetic science and how those powers have enriched our understanding of the roles they played in the modern history of Western civilization, both positive and negative. One of my human genetics seminars was centered on this volume and it was a remarkable experience for all.

Paul, who is currently Professor Emerita at the University of Massachusetts and a research associate at Harvard University, has now teamed with Jeffrey Brosco, a clinical pediatrician at the University of Miami School of Medicine, to explore the history of phenylketonuria (PKU), a rare but well-understood inherited metabolic disorder. PKU is the paradigm for successful medical intervention in genetic disease. If left untreated, a person born with PKU will develop debilitating intellectual disabilities. But if detected early and treated with a strict although difficult dietary regimen that restricts the intake of the amino acid phenylalanine, an essential amino acid, the prognosis for normal neurological functions is excellent.

The authors look at issues of patients living with PKU and look closely at the highly charged debates surrounding the problem. The narrative moves on to Ivar Asbjørn Følling’s landmark studies of the cause of PKU (Peter Harper includes Følling’s study in his collection, Landmarks in Medical Genetics: Classic Papers with Commentaries. 2004. Oxford (United Kingdom): Oxford University Press). Uncovering the metabolic basis of PKU led to strategies for trying to control the disorder by dietary intervention.

The final chapters examine new paradigms for PKU and look closely at the highly charged debates about the infamous genetics and IQ controversies of the late 1960s, 1970s, and 1980s (i.e., Arthur Jensen). The authors look at issues of patients living with PKU, the problem of maternal PKU, where a mother who has been successfully managed for PKU, then stops following the phenylalanine-restricted diet becomes pregnant with a non-PKU child who then becomes intellectually impaired because of the high levels of phenylalanine in the maternal circulatory system. They conclude with a thought-provoking discussion of political and eugenic issues.

Paul and Brosco discuss the fascinating development of mandatory testing for PKU as well as the skepticism that surrounded it. The PKU Paradox is strongly recommended to anyone who wants a wide-ranging, accessible account of the evidence that is being used to support the Hygiene Hypothesis. The caveat is that readers will need to do their own critical thinking and outside reading to adequately evaluate which alternative explanations are most compelling. Still, Velasquez-Manoff opens the door to considering the ways that the myriad residents of our bodies may influence our health, and the implications for reducing the damage that our immune systems sometimes inflict on us.

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