METABOLISM

FIRST SESSION

May 17, 1973, 9:00 A.M.

Continental Ballroom 4

MODERATOR: Michael M. Kaback

- 9:00 LETHAL NEONATAL HYPERAMMONEMIA SECONDARY TO CARBAMYL PHOSPHATE SYNTHETASE DEFICIENCY. Thomas D. Gelehrter and Philip J. Snodgrass, Yale Univ. Sch. of Med., Depts. of Human Genetics, Med. and Ped., New Haven, and Harvard Med. Sch., Dept. of Med., Boston (Intr. by Leon E. Rosenberg)
- 2. 9:15 ROLE OF CARNITINE IN SUBCUTANEOUS (WHITE) ADIPOSE TISSUE FROM HUMAN NEWBORN INFANTS. Milan Novak, Peter Hahn, Duna Penn, Ellen F. Monkus, and Josef P. Skala, Dept. of Ped., Univ. of Miami, Sch. of Med., Miami, and Dept. of Ped., & Obst. and Gynec., Univ. of British Columbia, Sch. of Med., Vancouver.
- 3. 9:30 AUTONOMIC AND ENZYMATIC CONTROL OF GLUCOSE PRODUCTION IN THE ISOLATED PERFUSED CANINE FETAL AND NEONATAL LIVER. Rowan Chlebowski, Satish Kalhan, Michael Lowry, and Peter Adam. Case Western Reserve University, Department of Pediatrics, Cleveland Metropolitan General Hospital.
- 4. 9:45 DEFECTIVE GLUCONEOGENESIS (GNG) IN SMALL FOR GESTATIONAL AGE INFANTS (SGAI) Morey Haymond, Irene Karl, Anthony Pagliara. Washington U. School of Med., St. Louis Children/s Hosp. Dept. Ped., St. Louis (Intr. Ralph Feigin)
- 5. 10:00 METABOLIC RESPONSE TO I.V. GLUCAGON IN SMALL FOR GESTATIONAL AGE INFANTS (SGA). D. Schiff, E. Colle, J. Aranda, A. Pappageorgiou, S. Reisner, C. Scriver and L. Stern. Dept. of Pediatrics, University of Alberta, Edmonton. McGill University-Montreal Children's Hospital Research Institute.
- 6. 10:15 STUDIES OF SERUM IMMUNOREACTIVE PARATHYROID HORMONE (IPTH) IN NORMAL AND HYPOCALCEMIC NEWBORN INFANTS. Louis David and Constantine Anast, Univ. of Missouri, Columbia.
- 7. 10:50 ISOVALERIC ACIDEMIA: A DEFECT OF LEUCINE METABOLISM IN CULTURED FIBROBLASTS. Kay Tanaka, Roseann Mandell and Vivian E. Shih. Depts. of Med. and Neurol., Harvard Med. Sch. and Mass. Gen. Hosp. Boston.

- 8. 11:05 ENZYME AND COENZYME FUNCTION IN MAPLE SYRUP URINE DISEASE, L. J. Elsas, F.B. Wheeler, B.A. Pask, J. H. Priest, Emory Univ. Sch. of Med., Dept. of Ped., Atlanta, Georgia, (Introduction by R.W. Blumberg).
- 9. 11:20 ABNORMAL LEUCINE/ISOLEUCINE RATIO AND THE ETIOLOGY OF ACRODERMATITIS ENTEROPATHICA-LIKE RASH IN MAPLE SYRUP URINE DISEASE (MSUD). John H. Diliberti, Angelo M. DiGeorge, and Victor H. Auerbach, Temple Univ. School of Medicine, St. Christopher's Hospital for Children, Dept. of Ped., Phila.
- 10. 11:35 EFFECT OF COPPER AND CERULOPLASMIN IN KINKY HAIR SYNDROME. William E. Bucknall, Neil A. Holtzman. The Johns Hopkins University School of Medicine, Department of Pediatrics, Baltimore.

METABOLISM

SECOND SESSION

May 19, 1973, 2:00 P.M.

Continental Ballroom 4

- 11. 2:00 TERMINATION OF PKU DIETARY THERAPY IN 62 PATIENTS, W. B. Hanley & L. Linsao (Intr. by Donald Fraser) Dept. of Paediatrics, Univ. of Toronto, Research Institute, Hosp. for Sick Children, Toronto.
- 12. 2:15 EVIDENCE FOR EPINEPHRINE DEFICIENCY AND ALTERED ALANINE AVAILABILITY IN "KETOTIC HYPOGLYCEMIA". Pierre C. Sizonenko, Luc Paunier, Michel B. Vallotton, Guy S. Cuendet and Errol B. Marliss (Intr. by Melvin M. Grumbach and Pierre E. Ferrier.) Univ. of Geneva Med. Sch., Hopital Cantonal, Dept of Ped., Geneva, Switzerland.
- 13. 2:30 PANCREATIC RESPONSE TO ACUTE AND CHRONIC GLUCOSE INFUSIONS IN OBESE CHILDREN. Robert H. Fiser, George A. Bray, & Delbert A. Fisher, Dept. of Ped. & Med., Harbor Gen. Hospital, UCLA School of Medicine, Torrance, Calif.
- 14. 2:45 TREATMENT OF PEDIATRIC FAMILIAL TYPES II AND IV HYPERLIPOPROTEINEMIA.
 C. J. Glueck, R. Fallat, R. Tsang. (Intr. by J. Sutherland). Univ. Cincinnati Coll. Medicine, Cincinnati General Hospital, GCRC and Dept. Pediatrics, Cincinnati.
- 15. 3:00 ANOREXIA NERVOSA: CLINICAL AND METABOLIC OBSERVATIONS. Joseph A. Silverman Columbia Univ. Col. of Physicians and Surgeons, Babies Hosp., Dept. of Ped., New York City. (Intr. by Robert W. Winters.)

- 3:15 Intermission -

- 16. 3:35 INDUCTION OF GAUCHER "LIKE" DISEASE IN NORMAL CULTURED FIBROBLASTS.
 Irwin A. Schafer, Norman S. Radin, Julia S. Sullivan, Mary Petrelli, and Kenneth
 Rabenowitz, Department of Pediatrics and Pathology, Case Western Reserve University
 and the Mental Health Research Institute, University of Michigan.
- 17. 3:50 THE SANFILIPPO A SYNDROME: A SULFAMIDASE DEFICIENCY. Reuben Matalon and Albert Dorfman, University of Chicago, Department of Pediatrics, Chicago.
- 18. 4:05 ENZYME THERAPY IN B-GLUCURONIDASE DEFICIENCY MUCOPOLYSACCHARIDOSIS. William S. Sly, Frederick E. Brot, Pra-on Chavalitdhamrong, and Philip D. Stahl, Washington Univ. Sch. Med., St. Louis Children's Hosp., Dept. Ped., Med., and Physiol. St. Louis.

275/47

- 4:20 EFFECT OF SERUM PROTEINS IN MPS II. N. DiFerrante, B. L. Nichols,
 G. Coppa and J. Singh. Baylor College of Medicine, Houston.
- 20. 4:35 FAILURE OF PLASMA INFUSION THERAPY IN MUCOPOLYSACCHARIDOSES. Maurice J. Mahoney and Y. Edward Hsia, Yale Univ., Depts. of Human Genetics and Pediatrics, New Haven. (Intr. by Charles D. Cook)