pounds, β-alanine, β-aminoisobutyric acid and taurine has been proposed on the basis of evidence found in the aminoacidopathy, hyper-β-alaninemia (New. Engl. J. Med. *274:* 635 [1966]). The proposal has been investigated further in the rat. Absorptive renal transport (lumen to cell) was selected for by using probenicid (200 mg/kg) to block tubular secretion of D-(-)βAIB. By means of intraperitoneal injection, the plasma concentration of β-amino compounds was raised; urinary excretion of amino acids was analyzed by chromatographic methods. Mutual competitive inhibition of absorptive transport was observed between βala, βAIB and Tau. βala had the greatest inhibitory effect, and Tau the least. β-amino compounds had no significant effect on the excretion of α-amino acids when the latter were present at either high or normal plasma concentrations; conversely, α-amino acids did not inhibit β-amino absorption. A common transport system for β-amino compounds operative at high concentrations, and whose order of substrate affinity is βala > L-βAIB> Tau, is thus demonstrable in mammalian kidney. (Supported by M.R.C. Grant, MA-1894, and N.I.H. Grant, AM-05117). (SPR)

50 *Bilirubin Nephropathy in the Gunn Rat.* GERARD B. ODELL, JURGEN C. NATZSCHKA* and G.N. BRUCE STOREY*, Dept. Pediatrics, Johns Hopkins Univ. Sch. Med., Baltimore, Md.

Homozygous, jaundiced, Gunn rats (jj) were compared with heterozygous control rats (jJ) for their capacity to concentrate their urine after water deprivation. Animals of comparable weights (5 jj and 6 jJ) were pair fed 7 days and then subjected to a 36-hour fast and thirst. Urine was collected for the last 6 hours, and the mean flow rates were 50 and 149 μl/100 g/h in the jJ and jj animals, respectively. The corresponding urine milli-osmolalities were 1909 and 815. Total solute loads excreted were comparable but the jj animals lost 3 times as much Na in the urine, and had a 30 % greater loss of body weight. Glomerular filtration rates were similar in hydrated jj and jJ animals. The concentrations of Na, K, Cl, NH_3 and urea in the renal cortex were similar in the two groups of animals. The concentrations of K and NH_3 of the medulla were also similar, but the concentrations of Na, Cl and urea in the medulla of the jj animals were only 1/3 that found in the jJ animals.

	Medullary analyses in mM/l tissue H_2O		
	Na	Cl	Urea
jj	103± 5.4 (S.E.)	91± 6.6	117±16.5
jJ	278±31.0	256±30.0	322±18.0

Regional analysis of the kidney for bilirubin demonstrated a 100 fold greater concentration in the renal medulla than in the corresponding cortex in jj animals. These results suggest that bilirubin may interfere with sodium and urea reabsorption in the medullary portions of the kidney and therby prevents the formation of hypertonic urine during thirsting comparable to normal rats. (SPR)

51 *Factor XIII—Report of a Family with Factor XIII Deficiency and the Concentrations in normal Infants.* JOHN D. BOUHASIN and CIGDEM ALTAY*, St. Louis Univ. Sch. of Med. and Cardinal Glennon Mem. Hosp., St. Louis, Mo. (introduced by Arthur E. McElfresh).

Since 1960 when DUCKERT *et al.*, observed a familial bleeding disorder due to a deficiency of Factor XIII, 21 cases, involving 8 families have been reported.

We have diagnosed Factor XIII deficiency in a 6-year-old boy with mild bleeding manifestations and studied the concentration of Factor XIII in his family and in normal infants.

An assay technique has been devised utilizing the patient's plasma as deficient substrate, with normal pooled plasma as the standard. Dilutions of plasma from 1:300 to 1:1000 yield a straight line with a steep slope when plotted on log-log paper against clot liquifaction time in minutes; it is reproducible. In vivo survival studies after transfusing the patient show a half-life of 5–7 days in agreement with other reports. Factor XIII was assayed in 50 infants and children from birth to 20 months of age as follows: newborn (10)-average 63 %, range 50–76 %; 0–5 months (10)-average 115 %, range 100–160 %; 6–9 months (10)-average 90 %, range 80–110 %; 10–15 months (10)-average 90 %, range 72–118 %; and 16–20 months (10)-average 94 %, range 70–120 %. Our adult range was 90 to 136 %. After the newborn period, Factor XIII is present in normal adult concentrations. Our data suggest a more rapid rise to normal and no evidence of the fall at ages 6–9 months as reported by KÜNZER (Ann. Pediat. *204:* 232 [1965]).

Family Factor XIII levels were: father 75 %, mother 48 %, 2 sisters 57 % and 48 % and brother 42 %. This tends to confirm the autosomal recessive inheritance of Factor XIII deficiency. (SPR)

52 *Effect of Diabetic Plasma in von Willebrand's Disease.* WM. E. HATHAWAY and H. GLEN HOSTETTER*, Univ. Colo. Med. Ctr., Denver, Colo.

Although in vivo correction of antihemophilic factor (AHF) levels is easily achieved in von Willebrand's disease (vWd), correction of the bleeding time (BT) defect is difficult by usual transfusion therapy. In an effort to find more effective treatment for four children with severe vWd (low AHF levels, prolonged BT, defective platelet adhesiveness), transfusion studies were done. Plasma AHF levels, BT's (modified Ivy), and native blood platelet adhesiveness tests were done before and after transfusions of fresh and fresh-frozen platelet-free ACD plasma. The results showed that plasma obtained from donors with diabetes mellitus (juvenile onset) corrected the bleeding time and platelet adhesiveness test temporarily in all four patients when doses of 10–15 ml/kg were used. Comparable dosages of normal plasma were effective in correcting the BT in only one patient. AHF-rich fibrinogen, cryoprecipitates of normal plasma, and plasma from an exercised donor did not correct the BT. The effectiveness of the diabetic plasma was approximately directly proportional to the severity of the diabetes. Also, the immediate rise in AHF levels was greater following diabetic plasma infusion than after normal plasma.

Mixtures of vWd blood and fractions of plasma were tested for platelet adhesiveness (PA) by the in vitro method of HELLEM. Diabetic plasma and cryoprecipitate showed excellent correction of the defective PA; AHF-fibrinogen and normal plasma showed moderate correction, and diabetic and normal serum, normal cryoprecipitate, and dextrose showed poor PA.

These in vitro and in vivo studies suggest that the blood from certain diabetics contains an increased amount of the factor(s) responsible for correction of