Homework: 1,3,4,5,6,9,11,17,19. **Introduction.** The amount of amino acid catabolism varies widely from species to species. Amino acids have to be broken down after 1) normal protein turnover, 2) a high protein meal or 3) in diabetes or starvation when the carbon is used for gluconeogenesis.

**18.1 Metabolic Fates of Amino Groups.** Amino acid catabolism involves removing nitrogen from the carbon chains, and then metabolizing the carbon. Glutamate and glutamine play an important role in this removal (Fig. 18-2). The removed nitrogen is simply excreted in ammonotelic organisms, and is excreted as a solid in uricotelic organisms. We are ureotelic and thus make urea. *Pyridoxal phosphate* (PLP) is very important in nitrogen metabolism, and you should be able to draw it (as in class) and understand mechanisms. See Fig 18-6 and online handout. In general most amino acids can transaminate, giving their nitrogens to $\alpha$-KG and becoming $\alpha$-keto-acids (Fig 18-4) with PLP as cofactor. Then glutamate DH can oxidatively remove the NH$_3$ (Fig 18-7). Thus $\alpha$-KG can carry one nitrogen (as Glu) or two nitrogens (as GIN) (Fig 18-8). Know the reactions of glutamine synthetase and *L*-glutaminase. Why does the body respond to metabolic acidosis (low level ketosis) with enhanced glutamine metabolism? (p. 681) Know the enzymes in Box 18-1 (mentioned in “Supersize Me”). Know the glucose-alanine cycle (Fig 18-9). Know that ammonia is toxic (p. 681).

**18.2 Nitrogen Excretion and the Urea Cycle.** Know everything about the Urea Cycle, including structures, enzymes, and cofactors. Know how *carbamoyl phosphate* is produced. Understand the "bicycle" that links the urea cycle to the citric acid cycle (Fig 18-12). Know the reactions of *N*-acetyl-glutamate synthetase and that the product, *N*-acetyl glutamate, stimulates CPS-I (Fig 18-13). Understand that the "urea bicycle" reduces the overall cost of urea synthesis from 4 ~P to 1.5 ~P (p. 686). Know the "ten" essential amino acids taught in class (refer to Table 18-1) and know the treatments for urea cycle deficiencies (p. 686-7).

**18.3 Pathways of Amino Acid Degradation.** Understand the concept of glucogenic and ketogenic amino acids (Fig 18-15). All amino acids with 6 or more contiguous carbons are at least partly ketogenic. *One carbon metabolism* (689-692) is extremely important and should be studied carefully. Be able to recognize THF structures, see Fig 18-17 and online handouts. Know structure and synthesis of *SAM* (Fig 18-18). Homocysteine is bad (not in book). Be able to draw *biotin* and know that B-12 or *cobalamin* is also a "one carbon cofactor."

*Pyruvate Family:* Know the lower four enzymes in Fig 18-19. *Acetyl CoA Family:* Know the first reaction in Fig 18-23 and the online handout about epinephrine synthesis (860). Understand PKU and *mixed function oxidases* (696-7).

*α–Ketoglutarate Family:* Know reactions in Fig 18-26 except His pathway.

*Succinyl CoA Family:* Know how Thr and homocysteine go to Succinyl CoA.

*Branched Chain Family:* Understand that these are metabolized in muscle, adipose, etc. by enzymes that resemble fatty acid beta oxidation enzymes. Lack of DH Complex leads to *Maple Syrup Urine Disease.* Read Box 18-2. Know AsN and Asp go to OAA.