

7.61 Discussion #D2 Membrane Proteins: Dystrophin

Assigned Reading for Discussion #2, Wed. September 20, 2006

COME TO THE DISCUSSION READY TO LEAD THE DISCUSSION. YOU SHOULD BE READY TO:

- 1) REVIEW THE SCIENTIFIC BACKGROUND UPON WHICH THE KEY QUESTION(S) ADDRESSED WERE BASED – WHY WAS THE WORK DONE? YOU SHOULD BE ABLE TO SUCCINCTLY DESCRIBE THE KEY QUESTIONS ADDRESSED IN THE PAPER
- 2) SUMMARIZE BRIEFLY EACH EXPERIMENT – FIGURE BY FIGURE AND TABLE BY TABLE -(IN ORDER OF PRESENTATION IN THE 'RESULTS' SECTION), INCLUDING:
WHAT QUESTION WAS ASKED, WHAT TECHNIQUE WAS USED AND WHY, HOW DOES THE TECHNIQUE WORK (E.G., SCANNING CALORIMETRY) AND WHAT CAN BE LEARNED BY USING THE TECHNIQUE, WHAT WERE THE RESULTS, WHAT CONCLUSIONS CAN BE DRAWN FROM THE RESULTS, DID THE AUTHORS DRAW REASONABLE CONCLUSIONS OR DID THEY GO TOO FAR OR NOT FAR ENOUGH IN INTERPRETING THE SIGNIFICANCE OF THEIR RESULTS
- 3) SUMMARIZE AND CRITICIZE THE DISCUSSION AND OVERALL CONCLUSIONS
- 4) PROVIDE A TWO OR THREE SENTENCE SUMMARY OF THE WHOLE PAPER

YOU SHOULD BE PREPARED TO GO TO THE BLACKBOARD TO CLEARLY SKETCH OUT ANY CONCEPT OR METHOD OR RESULT, IF THAT WILL HELP YOU PRESENT THIS INFORMATION SUCCINCTLY AND CLEARLY

BRING A FOLDED PIECE OF PAPER OR BOARD WITH YOUR NAME CLEARLY WRITTEN IN DARK, BOLD PRINT. THIS WILL SERVE AS A NAME PLATE FOR THE INSTRUCTORS AND YOUR FELLOW STUDENTS. PLEASE BRING THIS 'NAME PLATE' WITH YOU TO **ALL** DISCUSSIONS.

2 papers:

1. Ervasti, J.M. and Campbell, K.P. (1991). Membrane organization of the dystrophin-glycoprotein complex. *Cell* 66:1121-1131.
2. Ibraghimov-Beskrovnaya, O., Ervasti, J.M., Leveille, C.J., Slaughter, C.A., Sernett, S.W. and Campbell, K.P. (1992). Primary structure of dystrophin-associated glycoproteins linking dystrophin to the extracellular matrix. *Nature* 355:696-702.

For further background on muscular dystrophies, you may also want to look at the brief review Barresi R, Campbell KP. Dystroglycan: from biosynthesis to pathogenesis of human disease. *J Cell Sci.* 2006 Jan 15;119(Pt 2):199-207

Campbell, K.P. (1995) Three muscular dystrophies: loss of cytoskeleton-extracellular matrix linkage. *Cell* 80: 675-679.

Allamand V, Campbell KP. Animal models for muscular dystrophy: valuable tools for the development of therapies. *Hum Mol Genet.* 2000 Oct;9(16):2459-67.

Other papers on dystrophin:

Grewal PK, Hewitt JE. Glycosylation defects: a new mechanism for muscular dystrophy? *Hum Mol Genet.* 2003 Oct 15;12 Spec No 2:R259-64.

Barresi R, Michele DE, Kanagawa M, Harper HA, Dovico SA, Satz JS, Moore SA, Zhang W, Schachter H, Dumanski JP, Cohn RD, Nishino I, Campbell KP. LARGE can functionally bypass alpha-dystroglycan glycosylation defects in distinct congenital muscular dystrophies. *Nat Med.* 2004 Jul;10(7):696-703.

Bansal D, Campbell KP. Dysferlin and the plasma membrane repair in muscular dystrophy. *Trends Cell Biol.* 2004 Apr;14(4):206-13.

Smelt SC, Borrow P, Kunz S, Cao W, Tishon A, Lewicki H, Campbell KP, Oldstone MB. Differences in affinity of binding of lymphocytic choriomeningitis virus strains to the cellular receptor alpha-dystroglycan correlate with viral tropism and disease kinetics. *J Virol.* 2001 Jan;75(1):448-57.

Heathcote RD, Ekman JM, Campbell KP, Godfrey EW. Dystroglycan overexpression in vivo alters acetylcholine receptor aggregation at the neuromuscular junction. *Dev Biol.* 2000 Nov 15;227(2):595-605.